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Packing and Problematic Feeding Behaviors in CHARGE Syndrome: A Qualitative Analysis

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ABSTRACT

Objective: To understand packing and problematic feeding behaviors during mealtime in individuals with CHARGE syndrome. Packing, or holding food in one's cheeks without swallowing, is an adverse feeding behavior that has been described in children with autism and Down syndrome, and in those transitioning from tube to oral feeding. It has never been described in detail in CHARGE syndrome, a genetic disorder with a high prevalence of feeding difficulties, tube feeding, and otorhinolaryngological issues.

Methods: A mixed methods approach used descriptive and qualitative content analysis of interviews with parents of children, adolescents, and adults with CHARGE syndrome. Individuals had previously or were currently experiencing packing or overstuffing one's mouth with food during eating.

Results: Twenty parents completed a phone interview, describing their child/adult's (2–32 years) adverse feeding behaviors. Individuals had a higher proportion of cleft palates (40%) in comparison to the general CHARGE population (15–20%). Parents reported food packing most commonly with bread and pasta (33%), and reported that food was held in cheeks for hours after a meal had ended (35%). Packing was reported to prolong mealtimes for over an hour (30%). Parents were worried about choking during eating (30%). Food packing was also reported in individuals who had never needed G/J tube feeding or feeding therapy, in addition to those who had needed both.

Conclusion: This study provides an in-depth description of parents' experiences with packing and adverse feeding behaviors in individuals with CHARGE syndrome. These feeding behaviors are an important addition to the knowledge of the highly prevalent feeding difficulties in this genetic disorder. Individualized evaluation of feeding behavior should be a part of the standard otolaryngologic and feeding team practice for these patients.

1. Introduction

CHARGE syndrome is a genetic condition found in approximately 1 in 15,000 live births worldwide [1–4]. Diagnosis of CHARGE syndrome is initially clinical and confirmed by genetic testing [2,3]. Due to the broad range of clinical symptoms, extensive multidisciplinary treatment and management is needed, including assessment of all major organ systems and behavioral management [4]. Treatment and interventions can involve medical and surgical management including otolaryngology, deaf/blind services, occupational therapy, physiotherapy, speech/language therapy, cochlear implant, behavior therapy, psychological counseling, and more [4,5].

Children with CHARGE syndrome commonly experience feeding difficulties, with up to 90% reporting gastroesophageal reflux, poor sucking and swallowing skills, and G/J tube feeding [4,6–8]. Many parents report "odd" eating behaviors in their children with CHARGE syndrome when they transition from tube to oral feeding, such as taking large bites of food, overstuffing food into one's mouth before swallowing, and holding food in cheeks [6,7]. Severe feeding difficulties persist over the entire lifespan in this genetic condition, lasting throughout adolescence and adulthood [9–11].

Holding or pocketing food in ones' cheeks and mouth without swallowing has been described as 'packing' [12]. Packing has been identified as a problematic feeding behavior in children with autism spectrum disorders and children with feeding disorders [12–15]. Pocketing of food, or packing, has also been identified in Down Syndrome and described as leading to

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choking [16]. It has not previously been described in detail in CHARGE syndrome.

Packing, in addition to poor sucking and swallowing skills, may lead to an increased risk of aspiration, malnutrition, choking, and even death [16]. Aspiration and abnormal swallowing, including poor bolus mobility and pooling of secretions, have been previously identified in 60-80% of children with CHARGE syndrome [8]. Choking while eating has resulted in death in an eight-year-old child with CHARGE syndrome [17]. One of the authors (KB) is also aware of other individuals with CHARGE syndrome who had severe consequences due to choking on food. These consequences were anoxic cerebral palsy and a fatality due to choking on a piece of broccoli. Feeding difficulties, in addition to breathing difficulties and reflux, were significantly more prevalent in children with CHARGE syndrome who died before age 10 than those who survived longer [17].

Otorhinolaryngological and gastrointestinal tract surgeries are the most common surgeries in CHARGE syndrome [18]. Dental procedures and diagnostic scopes (nasopharyngoscopy, laryngoscopy, and bronchoscopy) are the next most common surgical procedures reported in children, adolescents, and adults with this genetic disorder. [18]. Excess oral secretions have also been identified as an issue in CHARGE syndrome [19]. Individuals may need to undergo botulinum toxin injections into their salivary glands to reduce oral secretions [19]. Numerous surgical procedures on the airway and gastrointestinal tract and excess salivary gland secretions can contribute to the feeding difficulties, dysphagia, and high risk of aspiration in this population [6,18,19]. ENT is one of the most common medical specialties that continues to follow individuals with CHARGE syndrome, with over 85% of adolescents and adults reporting ENT involvement in their care [9].

Despite the potential negative consequences of packing and the high prevalence of feeding difficulties in this genetic disorder, this problem has received little attention in the literature and has not been investigated in CHARGE syndrome. The objective of this study was to understand and describe packing and other problematic feeding behaviors in individuals with CHARGE syndrome through in-depth parent interviews. We aimed to understand the problematic feeding behaviors from the perspective of the parents themselves.

2. Methods

2.1. Demographics survey

Information regarding demographics as well as the major and minor phenotypic features of CHARGE syndrome was collected (Appendix A) [5,20].

2.2. Semi-structured interview

Participants completed a semi-structured interview consisting of open-ended and closed-ended questions (Appendix B). The interview questions explored general eating, problematic feeding behaviors (e.g., packing), chewing, swallowing, choking, and feeding therapy. The interview questions for the semi-structured interview were developed from the current literature on food packing [11,17–19], and feeding difficulties experienced in CHARGE syndrome [6,8,12]. Several interview questions were based on the Pediatric Assessment Scale for Severe Feeding Problems [20]. Clinicians and experts in pediatric feeding issues reviewed and revised the interview questions. Reviewers included a general pediatrician whose clinic includes individuals with CHARGE syndrome, a speech language pathologist specializing in feeding, a feeding specialist from a pediatric feeding team, an occupational therapist, a pediatric gastroenterologist, and a parent of an older child with CHARGE syndrome.

2.3. Participants

Parents of individuals with CHARGE syndrome who were currently experiencing or had previously experienced food packing, overstuffing one's mouth during eating, or both feeding behaviors, were included in this study. Individuals had to have a confirmed genetic or clinical diagnosis of CHARGE syndrome and be older than 1 year.

2.4. Procedure

Approval for this study was obtained through the I.W.K. Health Centre Research Ethics Board. Invitations to participate were sent via CHARGE syndrome organizations in the USA, Canada, Australia, and the UK. Members of these international and national organizations included parents of individuals with CHARGE syndrome. The invitations included information about the study and eligibility criteria.

Informed consent was obtained from each parent before participation. Participants completed the Demographic survey and the pre-set interview questions during a recorded phone interview with the primary author [AH]. Interviews were conducted until data saturation was reached for the pre-set questions. The primary author [AH] conducted all of the interviews. Interviews were audio recorded and transcribed verbatim. The transcriptions were checked for accuracy.

2.5. Analysis

Interviews were analyzed using a qualitative content analysis of open-ended questions and a descriptive analysis of closed-ended questions. Continuous variables were expressed as mean (M)±standard deviation (SD) and categorical variables were expressed as frequency (N) and percentage (%). All analyses were done in SPSS Version 20.0.0 (IBM® SPSS© Statistics, 1 New Orchard Road Armonk, New York).

The primary author reviewed the responses to the open-ended questions and created content categories, with definitions for each category. The data was coded by these categories. The number of responses in each category was summed. Separate categories were created for each of the open-ended questions. All analyses were done in MAXQDA Version 11.2.1 (Udo Kuckartz Berlin©). Direct quotations from the interviews were taken to support the categories. A second coder independently reviewed and validated the coding and content categories.

3. Results

3.1. Participants

Between April and August 2015, 20 parents (95% mothers) of individuals with CHARGE syndrome (12 female, 60%) completed a phone interview. Interviews took an average of 44 min (SD = 11 min). Fifteen individuals (75%) had a confirmed genetic diagnosis of CHARGE syndrome (CHD7 mutation positive). One individual (5%) did not demonstrate the CHD7 gene mutation and four individuals (20%) had not undergone genetic testing, but all clinically met the criteria for typical/definite CHARGE syndrome using both the Verloes criteria (2005) [15] and the Blake criteria (1998) [5].

The average age of individuals with CHARGE syndrome was 9.9 years (SD = 8.6 years) and ranged from 2 to 32 years. Individuals were from USA (n = 11, 55%), Canada (n = 4, 20%), Australia/New