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TASTE PERCEPTION AND SENSORY SENSITIVITY: RELATIONSHIP TO FEEDING PROBLEMS IN BOYS WITH BARTH SYNDROME

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ABSTRACT

Background: Feeding problems are common in boys with Barth syndrome and may contribute to the population's propensity for growth delay and muscle weakness.

Objectives: The purpose of this study was to quantify and describe feeding issues in Barth syndrome and examine altered taste perception and sensory sensitivity as contributing factors.

Methods: A cross-sectional, 2-group comparison design was used to examine feeding preferences and behaviors, chemical taste perception, and sensory sensitivities in 50 boys aged 4 to 17 years with (n = 24) and without (n = 26) Barth. Taste perception was measured using chemical test strips saturated with phenylthiocarbamide (PTC) and sodium benzoate (NaB). Feeding problems were documented by parents using a food inventory, and sensory sensitivities were recorded using the Short Sensory Profile.

Results: Boys with Barth differed significantly from typical peers with regard to problem feeding behaviors. Food refusal and food selectivity were identified as being present in ~50% of the sample, while 70.8% had identified problems related to gagging or swallowing foods. About half of all Barth families noted that their child's eating habits did not match the family's and that separate meals were often prepared. About 50% of the boys demonstrated probable or definite differences in taste/smell sensitivity, which was significantly higher than in controls. On tests of chemical taste perception, boys with Barth were significantly more likely to be supertasters to PTC ($P < .05$) and nontasters to NaB ($P < .01$).

Taster status did not directly relate to the presence of feeding problems; however, taste/smell sensitivity did significantly relate to food selectivity by type and texture.

Conclusions: Feeding problems were found in at least 50% to 70% of these boys with Barth syndrome, and were often present before 6 months of age. Differences in taste perception may influence dietary choices in boys with Barth, particularly their craving of salty foods. Taste/smell sensitivity also appears to influence food selectivity and therefore may be important to consider in this population, particularly in light of dietary influences on cardiac function, energy consumption, and overall growth.

INTRODUCTION

Barth syndrome is a rare X-linked genetic disorder with ~150 living cases and 500 known cases identified in the Barth Syndrome Registry.[1] It is a potentially fatal disease caused by mutation of the tafazzin (TAZ) gene on chromosome Xq28, resulting in a loss of function in the protein product tafazzin, a transacylase. The alteration in tafazzin activity results in remolding of cardiolipin in mitochondrial membranes. Structural and functional changes in mitochondrial integrity are believed to be primarily responsible for the Barth phenotype, characterized by core features of cardiomyopathy, skeletal myopathy, and neutropenia.[2] Other common features of the disorder include feeding problems, exercise intolerance, and delays in growth, gross motor milestones, and puberty. Of these characteristics, the issue of feeding problems has received little attention in the literature, although it is commonly discussed among those familiar with the disorder.

Anecdotally, many parents report that, in infancy, their child with Barth had difficulty transitioning to solid foods and gagged frequently during mealtimes or even in the presence of food. In a report documenting 4 cases of Barth in the Czech Republic, feeding problems were present in 3 of the boys at the time of their initial referral at 3 to 4 months of age.[3] Data from the Barth Syndrome Registry suggest that approximately a third of boys with Barth require a nasogastric or gastrostomy tube for feeding at some point.[4] These feeding problems have the potential to contribute not only to growth delay and muscle weakness, but also may add to family stress, with parents concerned about their child's nutritional intake and troublesome mealtime behaviors.

Unfortunately, feeding problems and mealtime behaviors in boys with Barth syndrome do not appear to resolve after infancy. In a study published in 2012, our team used qualitative methods to examine feeding issues in boys with Barth aged 4 to 17 years.[5] These boys were reported to have a restricted repertoire of foods they would eat, with many continuing to exhibit an abnormally sensitive gag reflex. Interestingly, boys with Barth were also identified as having very strong taste preferences, with most boys preferring foods that were very salty, cheesy, or spicy. The study concluded that atypical taste/smell and tactile sensitivities may heavily contribute to feeding problems in the Barth population, and that this was an area in need of further research.

The purpose of the present study was to (1) quantify and compare the presence of atypical feeding behaviors in boys with and without Barth syndrome, (2) quantify and compare chemical taste perception and other sensory sensitivities in boys with and without Barth, and (3) examine the relationship between problematic feeding behaviors, taste perception, and sensory sensitivities in boys with Barth. Our primary hypothesis was that boys with Barth would be more likely to be classified as "supertasters" with regard to bitter chemical taste sensitivity, and secondly, that taste perception would influence food refusal and food selectivity.

Taster Perception and Food Selection

Taste perception is based on the chemical sensitivity of receptors located on the tongue; genetic variation in taste-receptor sensitivity generates unique perceptions of certain tastes.[6] It has been shown that, at least to some extent, taste perception influences food preferences and, along with environmental influences, may contribute to dietary choices.[6,7] The most well-studied chemical taste receptor gene is TAS2R38, which encodes for bitter receptors detecting the thiourea compounds phenyltiocarbamide (PTC) and 6-n-propylthiouracil (PROP). While there is a natural range in which these bitter compounds are detected, individuals who perceive PTC/PROP to be intensely bitter have sometimes been termed "supertasters."[8] Over the past 2 decades, research has suggested that individuals who have a higher sensitivity for these bitter compounds may report less liking of vegetables and consume fewer vegetables overall [7,9,10]; they also may consume less fruits compared with low-sensitivity tasters.[11] Alternatively, nontasters, those who least taste the bitterness in PROP, have reported consuming more alcohol and have a greater preference for high-fat and sweet foods.[12]

Markedly less studied than bitter taste, salty-taste perception has been examined and found to have a relationship to perception of bitter tastes. Perceived bitterness of PROP has been associated with a higher perceived intensity of aqueous salt samples[13] and perceived saltiness in foods.[14] PROP supertasters, in one study, were shown to be most sensitive to sodium changes in chicken broth, and disliked broths with the highest sodium concentration.[14] While PROP supertasters did show frequent high-sodium food intake, researchers suggested that in the contemporary salt-rich American diet, supertasters may find foods to be sufficiently salty, while nontasters may seek to increase the stimulus intensity to achieve desired levels of saltiness.

Given the potential influence of taster status (eg, supertaster vs nontaster) on dietary food choices, these phenotypic behaviors may be important to consider with relation to Barth syndrome, particularly in light of dietary influences on cardiac function, energy consumption, and immunologic functions.

Sensory Sensitivities and Feeding Behaviors

In populations with childhood genetic disorders, no studies to date have correlated sensory sensitivity with problematic feeding behaviors, such as food selectivity. However, studies on typical children have associated selective eating with sensory sensitivity in tactile and taste/smell domains. In a study by Farrow and Coulthard,[15] taste/smell and tactile sensitivities were associated with a child's lower consumption of fruits and vegetables, and with higher food neophobia.[16] Our previous research found that ~50% of children with Barth syndrome had a probable or definite difference in taste/smell sensitivity.[5] Therefore, taste/smell sensitivity could account for the food selectivity seen in this population.

METHODS

Design

A cross-sectional, descriptive, 2-group comparison design was used. All procedures were approved by the universities' internal review boards prior to subject recruitment. Consent and assent procedures were completed for all participants.

Subjects

Twenty-five males with a diagnosis of Barth syndrome between the ages of 3 and 17 years were recruited at the Barth Syndrome Foundation Conference held in St. Petersburg, Florida, in June 2012. A convenience sample of 25 typical boys ages 3 to 17 years with no known medical (eg, heart condition, cancer), genetic (eg, fragile X or Angelman syndrome) or psychological diagnoses (eg, autism, bipolar) were recruited from community centers, schools, and youth groups in an urban university setting. All boys and their caregivers demonstrated the proficiency in English needed to follow instructions and complete written forms.

Tools

Food Inventory

Parents of boys with Barth syndrome and control subjects were asked to complete a 4-part food inventory questionnaire. The first section asked questions about unusual or problem behaviors associated with food intake or food preference. In response to each behavior or symptom listed, parents responded "yes" or "no"; if parents responded "yes," they were asked to write details about the behavior, including age of onset. The second section of the questionnaire asked parents to provide information about the frequency that specific foods were eaten by the child and if those foods were eaten by the family. Drinking and meal patterns were queried in the third section, which included questions such as "Does your child often complain of being thirsty?", "Do your child's food habits match the family's?", and "How would you describe your child's appetite?" Similar to the first section, parents were asked to provide written details about unusual behaviors. In the fourth section, parents were asked to complete a checklist about whether or not the child was able to eat specific food consistencies (eg, smooth or creamy foods, chewy foods); response options included that the child "can eat" the food, "won't eat" the food, or "has never tried" the food.

Evaluation of Taste Perception

To measure taster perception, subjects were asked to rate responsiveness to PTC and sodium benzoate (NaB, a salty compound) using standardized test strips (Indigo Instruments, Tonawanda, NY). All boys with Barth were tested over a period of 3 days at the Barth Syndrome Foundation Conference. Boys were accompanied by parents to a conference room in the hotel and completed all testing procedures with minimal distractions. Testing of control boys was conducted at a location convenient for families, which included the family home, the child's school, or a community center. In all cases, researchers attempted to limit outside distractions during taste testing.

The taste-test protocol was administered in the same way to both Barth and control groups. Procedures for the taste test were first explained to the child and family, and questions were answered. Procedures began by asking the children to rinse their mouth with room-temperature water and spit the water into a disposable cup. Next, the PTC strip was placed lengthwise onto the child's tongue; the child was then asked to close his mouth and let the strip rest on his tongue for 10 seconds. After removing the strip, the child was asked to rate the taste by pointing his finger along a visual analog scale ranging from "no taste" to "intensely bitter or revolting." Subjects then repeated the water-rinsing procedure at least one time, or until they reported no aftertastes of the test strip. The entire taste-test procedure was repeated for the NaB test strips. Taste-test answers were marked on the visual analog scale and were later converted to a scale score ranging from 1 to 5, using an overlay sheet with the numeric scale. Children with scores ranging from 1 to 1.5 were categorized as nontasters, scores of 1.6 to 4.4 were categorized as moderate tasters, and scores of 4.5 to 5 were categorized as supertasters.

Short Sensory Profile

The Short Sensory Profile (SSP) is a 38-item questionnaire that examines a child's behavioral reactions to various sensory situations found in everyday life.[17] Parents completed the SSP for all participants, ranking each behavior on a frequency scale ranging from "always" (child always responds in this manner) to "never" (child never responds in this manner). For the purposes of this study, only section scores related to taste/smell sensitivity and tactile sensitivity were included, as these were the sections most relevant to feeding. For each section, the child's raw score was recorded and then classified into 1 of 3 categories: typical performance (at or above 1 standard deviation [SD] below the mean), probable difference (between 1 and 2 SDs below the mean), and definite difference (more than 2 SDs below the mean).

Statistical Analysis

All data were entered into the SPSS 21 statistical analysis program. The first 2 study objectives involved comparison of boys with and without Barth syndrome.

Descriptive statistics were initially run for both groups. Pearson χ^2 tests were used to compare boys with and without Barth for all categorical dependent variables (food inventory, taste perception, taste status). A one-way analysis of variance was conducted to compare mean scores (continuous raw scores) on the SSP for boys with and without Barth. Parental written responses from the food inventory were transcribed into a separate word processing document; specific trends (eg, number of times gag reported) were counted and are reported as descriptive data.

The third objective of the study was to test whether taste perception and/or behavioral report of taste/smell sensitivity are related to the presence of atypical food behaviors in boys with Barth syndrome. A Pearson X2 test was used to examine the relationship between taster status (nontaster, moderate taster, and supertaster) and the presence of salient problem behaviors. A similar analysis was run to examine the relationship between categories of taste/smell performance (typical, probable difference, and definite difference) and the presence of salient problem behaviors. The alpha level for all statistical tests was set at 0.05.

RESULTS

Sample Characteristics

Of the 25 boys with Barth syndrome recruited for this study, only one child (a 3-year-old) was unable to complete the protocol. The remaining 24 boys in the Barth sample ranged in age from 4 to 17 years with a mean (SD) age of 9.92 (4.3) years. Owing to an aggressive sample strategy, data were collected on 26 typical boys instead of the targeted 25. Because our target N was 50 for the study, it was decided that all data would be included in the final analysis. Boys in the typical sample ranged in age from 5 to 17 years, with a mean age of 11.7 (3.6) years. Although the mean age for the Barth sample was lower than that of the control group, these differences were not significant (P = .106).

Of the boys in the Barth group, 28% were born prematurely, compared with only 3.8% of boys in the control group. Food allergies were more common in the control group, as were seasonal allergies. Presence of attention deficit hyperactivity disorder (ADHD), learning disabilities, and asthma were similar between groups. A full description of group characteristics can be found in **Table 1**.

Food Behaviors and Preferences

A 2×2 Pearson χ^2 test of independence was used to examine the relationship between Barth syndrome and behaviors listed on the food inventory. The grouping variable was diagnostic group (Barth, control), and the outcome variable was presence of feeding behavior (yes, no). The relationship between all examined variables was significant (**Table 2**).

Very few children in the control population were noted to have problem behaviors related to feeding or eating.

Table 1. Participant Diagnostic Characteristics (N = 24)

	Never, %		Previous, %		Current, %	
	Barth	Control	Barth	Control	Barth	Control
ADHD	87.5	80.8	-	-	12.5	19.2
Learning disability	95.8	100	-	-	4.2	-
Asthma	95.8	92.3	-	-	4.2	7.7
Food allergies	95.8	80.0	-	-	4.2	20.0
Seasonal allergies	70.8	65.4	8.3	-	20.8	34.6
Diabetes	95.8	100	-	-	4.2	-
Hearing impairment	95.7	100	4.3	-	-	-
Reflux	87.5	96.2	4.2	3.8	8.3	-
Nasogastric intubation	66.7	100	33.3	-	0	-
Nasal cannula	87.5	100	12.5	-	0	-
Gastrostomy tube	95.8	100	4.2	-	0	-

Three respondents noted that their typical child refused to eat fish, while 2 reported that their child would not eat mashed potatoes. One child in the control group was noted to have a strong aversion to green vegetables, while another had a strong preference for cereal and pasta. Only one 5- year-old child in the control group was noted to have problems with dysphagia (ie, problems swallowing and gagging in response to food), whereby the child would immediately cough and spit out any food he didn't like.

In contrast, dysphagia was the most prevalent behavior noted in the Barth population (70.8 %). Eight caregivers reported that these problems were present at birth or during early infancy. While 12 of the 17 caregivers indicated problems with hypersensitive gag (elicited by actual foods and food smells), 3 respondents noted problems with the actual mechanisms of swallowing.

Half of the Barth population (50%) were also reported to engage in food refusal behaviors, defined as refusing all or most food. Of the 12 caregivers, 7 noted that food refusal started at birth or during infancy. Two caregivers noted that their child outgrew this behavior; one family noted the behavior improved with time. Selectivity based on food type (eating only a narrow variety of foods) also began for most boys with Barth syndrome in infancy around the time they began to eat solid foods. Of the 13 boys with Barth (54.2%) who were reported to have food selectivity by type, 4 were identified as having a strong preference for salty foods and 3 were reported to strongly prefer cheese or dairy items. Food selectivity by texture was defined as only certain textures or refusing to eat certain textures. Nine respondents noted that problems tolerating food textures started at birth or when their child was first introduced to solid food. When more specific information was gathered about aversion to food textures, parents identified mashed table foods (eg, mashed potatoes) and soups with pieces of meat or vegetables as the foods their children with Barth wouldn't eat or have never tried (**Table 3**).

Unusual food preferences and behaviors were also present in about a third of the Barth sample. Unusual food preferences was defined as eating only certain brands of foods, refusing to eat a preferred food if it was not a specific temperature, or needing a certain cup or preferred utensil to eat. Unusual feeding behaviors was defined as other behaviors or attitudes regarding eating or drinking that the parent or caregiver believed were atypical. Only 4 families provided a written response for these items. Two families noted temperature preferences, with one of these families indicating that their child was very specific about the temperature of his bottle. One additional family reported that their child

Table 2. Food Behaviors and χ^2 Analysis

	Barth, % yes	Control, % yes	χ^2 Value	*df* (n =)	*P* Value
Food refusal	50.0	15.4	6.872	1 (50)	.009 *
Selectivity by texture	50.0	15.4	6.872	1 (50)	.009*
Selectivity by type	54.2	11.5	10.422	1 (50)	.001*
Dysphagia or gag	70.8	3.8	24.036	1 (50)	<.001*
Unusual food preferences	37.5	3.8	8.834	1 (50)	.003*
Unusual food behaviors	33.3	7.7	5.128	1 (50)	.024*
Complain of being thirsty	30.4	0	9.232	1 (49)	.002*
Poor appetite	23.8	3.8	4.157	1 (50)	.041*
Eating habits match the family	43.5	84.6	9.115	1 (50)	.003*
Requires separate meals	58.3	23.1	6.464	1 (50)	.011*

*Indicates statistically significant differences between groups.

Table 3. Food Consistency Preferences

Texture	Can Eat, %		Won't Eat, %		Never Tried, %	
	Barth	Control	Barth	Control	Barth	Control
Soups	69.6	92.3	26.1	7.7	4.3	-
Creamy foods	95.7	100	4.3	-	-	-
Mashed food	70.8	84.6	25.0	15.4	4.2	-
Crisp food	100	100	-	-	-	-
Chewy food	91.7	100	8.3	-	-	-
Crunchy food	95.8	92.6	4.2	3.8	-	

Table 4. Taster-Status Outcomes

	Nontaster, %		Moderate Taster, %		Supertaster, %	
	Barth	Control	Barth	Control	Barth	Control
PTC	40.9	44.0	27.3	52.0	31.8	4.0
NaB	63.6	20.0	36.4	72.0	0	8.0

NaB, sodium benzoate; PTC, phenylthiocarbamide.

Table 5. Reported Outcomes on the Short Sensory Profile

	Mean (SD)		Typical Performance, %		Probable Difference, %		Definite Difference, %	
	Barth	Control	Barth	Con-trol	Barth	Control	Barth	Control
Tactile sensitivity	30.1 (4.1)	31.2 (3.4)	65.2	73.1	17.4	19.2	17.4	7.7
Taste/smell sensitivity	13.8 (5.5)	17.3 (3.7)	50.0	80.8	12.5	11.5	37.5	7.7

SD, standard deviation.

required the use of separate utensils for each food eaten during the meal, and foods could not touch on the plate. Two respondents noted that their child ate very small meals or "grazed" throughout the day as opposed to eating at mealtimes. This is commensurate with the finding that nearly a quarter of boys with Barth (23.8%) in our sample have a small appetite.

When asked about whether their child's food habits and preferences matched the family's, more than half of Barth families (56.5%) reported "no." Similarly, about half of Barth families acknowledged that it was common for caregivers to prepare a separate meal for their child because he would not eat the family meal.

Taste Perception

A 2×3 χ^2 test of independence was performed to examine the relationship between Barth syndrome (grouping variable = Barth, typical) and taster status to PTC (outcome variable = nontaster, moderate taster, supertaster). The relationship between these variables was significant: χ^2 (2, N = 47) = 7.116; $P < .05$. Boys with Barth were more likely to be supertasters to PTC compared with boys without Barth. Boys in the control group were more likely to be moderate tasters . A similar analysis was performed to examine the relationship between Barth and taster status to NaB. The relationship between these variables was significant: χ^2 (2, N = 47) = 9.958; $P < .01$. Boys with Barth were more likely to be nontasters to the salty NaB compared with boys without Barth. Boys in the control group were more likely than boys in the Barth group to be moderate tasters or supertasters. Percentages for each group are shown in **Table 4.**

Behavioral Responses to Sensation

A one-way analysis of variance revealed significant differences between the groups ($F(1,49) = 7.178$; $P < .05$) for the taste/smell sensitivity variable. No group differences were found in the area of tactile sensitivity ($F (1,48) = 1.131$; $P = .293$). The means and SDs are presented in **Table 5**, along with the corresponding categories of performance.

Relationship Between Taste Perception, Sensory Sensitivity, and Food Behaviors

Our X2 test revealed that, in boys with Barth syndrome, the presence of atypical food behaviors did not significantly differ based on PTC or NaB taster status. Boys with Barth who had definite differences in taste/smell sensitivity were more likely to be more selective eaters, both by food texture (χ^2 (2, N = 24) = 6.111; $P < .05$.) and food type (χ^2 (2, N = 24) = 8.671; $P < .05$). They also tended to have more unusual food-related preferences, though this finding was not significant (χ^2 (2, N = 24) = 5.511; $P = .064$).

DISCUSSION

The aim of this study was to quantify and describe feeding issues, taste perception, and sensory sensitivities in boys with and without Barth syndrome, as well as examine the relationship between these factors. The primary hypothesis, that boys with Barth would be more likely to be classified as PTC supertasters, was based on our previous study, in which parents had identified a high rate of food refusal (particularly vegetables) and food selectivity in this population.[5] Taste perception research has suggested that PTC supertasters have a lower intake of vegetables and citrus fruits, as well as a greater fear of trying new foods and more disliked foods.[18] Findings from the current study supported our hypothesis, with approximately a third of boys with Barth being identified as supertasters compared with only 4.0% of the control sample. Interestingly, boys with Barth were also found to differ from typically developing peers in the perception of NaB. In this instance, boys with Barth were more likely to be nontasters as opposed to supertasters, suggesting a diminished threshold for salty tastes. At first, this finding may appear to be in contrast with prior reports that boys with Barth syndrome crave salty foods (eg, snack chips and processed cheese products) and add liberal amounts of salt to already salted foods such as French fries.[5] However, it has been suggested that while supertasters may be more capable of detecting and discriminating salt in foods, nontasters may seek to increase the stimulus levels to achieve desired hedonic levels.[14] In this way, our findings of PTC supertaster status and NaB nontaster status in boys with Barth align well with the dietary profile of Barth children, which includes a strong preference for salty/cheesy foods and an overall picky eating profile.

Our second study hypothesis, that taste perception would influence feeding problems such as food refusal and food selectivity, was not supported. Our findings indicate that taster status (nontaster, moderate taster, or supertaster) did not significantly relate to the presence or absence of any feeding problems outlined in the food inventory. This could be due to several factors. First, while taster perception has been strongly linked to dietary preferences, it is likely only one of several complex factors that lead to childhood feeding problems. One factor to consider may be the general medical complications associated with Barth syndrome, including prematurity and cardiac disease, which could result in early struggles with caloric intake. Commensurate with previous reports, about a third of our sample (33.3%) had a history of nasogastric intubation, while another 4.2% had received a gastrostomy tube. Although tube feeding is commonly used to ensure appropriate nutrition and growth in sick children, those who are tube fed often have difficulty transitioning to oral feeding and may develop sensory sensitivities that limit the range of tastes and textures they tolerate in their mouth.[19] Nasogastric tubes may also cause gastroesophageal reflux, a condition characterized by chest pain and nausea, which was reported to occur in 12.5% of our Barth sample. Mason and colleagues acknowledge that exposure to a food, followed by nausea, pain, or vomiting, can cause an immediate link between that food and the averse experience, leading to dislike and avoidance of that food.[19] Therefore, early averse experience with feeding may lead to behavioral patterns seen later in the form of food refusal, food selectivity, or unusual food behaviors. Lack of oral feeding may also limit a child's ability to inhibit the naturally strong gag reflex present at birth, which usually diminishes (or moves to the more posterior part of the tongue) by 7 months of age. Gagging was commonly reported in our Barth sample (~70%), and may interfere directly with the development of food preferences and willingness to try new foods.

In addition to general medical concerns, other symptoms of Barth syndrome may influence food selectivity or refusal. Muscle weakness and lack of energy may influence chewing skills or the ability to manipulate food inside the mouth; this could lead to an avoidance of certain food textures that is more difficult to manage. Food textures that were most likely to be refused by our sample of boys with Barth were soups with pieces of meat or vegetables and mashed table foods that are often inconsistent in texture. While tactile hypersensitivity was not significantly different between boys with and without Barth, approximately 35% of the Barth sample were within the probable or definite difference category for this sensory feature. Thus, there is not sufficient evidence to rule out the influence of texture and tactile sensitivity on feeding behaviors in this population."

The strongest relationship between sensory processing and feeding problems in boys with Barth syndrome was in the area of overall taste/smell sensitivity. This is particularly interesting in light of the finding that taste perception did not significantly influence feeding behaviors. One potential reason for this discrepancy is the inclusion of smell (olfaction) as a sensation influencing food selection. Olfaction is a known contributor to flavor perception and dietary choices.[8] Olfaction also influences an individual's hedonic evaluation of food and has been shown to directly influence one's willingness to try new foods.[20] Similar to tastes, smells can also elicit gagging and nausea and can be used to link certain foods to averse experiences in one's memory. This study did not include an evaluation of olfactory threshold or identification abilities, although this would certainly be an area for future study in the Barth population.

Study Limitations

A primary limitation of this study was the use of parent report for information related to feeding behaviors and sensory sensitivities. In many cases, parents estimated the age at which they remembered behaviors occurring or initiating, which may have been distorted by time. While our sample size was relatively small, it reflects an admirable percentage of boys living with this rare disorder (~16 %).

CONCLUSIONS

The high percentage of feeding problems identified in boys with Barth syndrome suggests that this is a primary phenotypic behavior associated with this rare genetic disorder. Owing to the early onset of these behaviors, clinicians should be prepared to support families during the early years, when oral feeding is initiated and a critical period for the development of oral motor skills. This will likely require the use of a multidisciplinary team approach that includes medicine, nursing, nutritional services, and occupational and physical therapy.

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