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A Mixed-methods Investigation of Sensory Response Patterns in Barth Syndrome: A Clinical Phenotype?

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Abstract

Barth syndrome is a genetic disorder that affects approximately 1/500,000 boys each year. While treatment of medical complications associated with Barth is of primary importance, there is a concomitant need to look at behavioral and clinical features of the disorder. The purpose of this study was to examine the prevalence of atypical sensory processing in 21 boys with Barth syndrome and to explore if phenotypic patterns of sensory responsiveness may be useful in early diagnosis. Using a mixed methods approach, we found that sensory issues related to feeding and eating were ubiquitous in our sample, with some behaviors such as strong gag reflex identifiable early in development. Specifically, boys with Barth had a strong preference for salty, cheesy, and spicy foods while having an overall restricted repertoire of foods they would eat (e.g. picky eaters). In boys with Barth as they age, auditory sensitivity and auditory filtering issues also emerged as potential sensory-related behaviors affecting academic performance and participation. Overall, this study suggests that early identification of sensory patterns in Barth may assist in differential diagnosis and create opportunities for early interventions that may minimize the impact of these behaviors on function and participation.

Keywords

Barth syndrome; Sensory Processing; TAZ gene; cardiomyopathy; neutropenia; Sensory Profile

INTRODUCTION

Barth syndrome is a genetic disorder characterized by cardiomyopathy and neutropenia with other associated features such as growth retardation and hypocholesterolemia [Spencer et al., 2006]. Since it is an X-linked disorder, Barth syndrome affects only boys; however, the general manifestation of the disorder appears to vary between affected males and within the individual throughout the lifespan. While strides have been made to control the immediate medical complications associated with Barth syndrome, there is a need to focus on better understanding the behavioral phenotype associated with the condition, both for early diagnosis and for purposes of therapeutic intervention. The purpose of this study was to examine behaviors associated with sensory responsiveness in males aged 4–17 diagnosed with Barth syndrome. A mixed methods approach was used to assess whether unique patterns of atypical sensory behaviors are present in individuals with Barth syndrome and

explore whether these behaviors may be considered phenotypic features specific to this genetic disorder.

Sensory responsivity

The ability to register and process sensory information from the environment is one of the earliest tasks the human nervous system is capable of, and forms the foundation for future higher level cognitive, motor, and social emotional abilities [Ayres, 1998]. It is believed that disruptions in the ability of the nervous system to process sensation can lead to observable behavioral responses that are atypical in nature [Miller et al., 2007].

Atypical responses to sensory stimuli have been documented in childhood genetic disorders such as fragile X syndrome, Rett syndrome, and Angelman syndrome [Baranek et al., 2008; Mount et al., 2003; Walz and Baranek, 2006]. These atypical responses have been reported across sensory modalities (e.g. tactile, auditory, gustatory), with varying severity and consistency across the lifespan. Both *under-responsiveness* (i.e. failure to notice or respond to stimuli) and *over-responsiveness* (i.e. exaggerated responses to or avoidance of stimuli) have been documented, as well as behaviors associated with *sensation seeking or craving*. Collectively, sensory under-responsivity, sensory over-responsivity, and sensation seeking are believed to constitute a condition known as sensory modulation disorder (SMD), a disorder in which the individual is unable to match behavioral responses to the demands of the environment [Miller et al., 2007; Lane, 2002]. While the underlying neurobiology of SMD is currently unknown, physiological and neuroendocrine data support behavioral reports associated with exaggerated or diminished sensory responsiveness in certain populations of children with neurodevelopmental disorders [Reynolds et al., 2010; Schaaf et al., 2010; Schoen et al., 2009].

While many children and adults have isolated sensory preferences that may be considered atypical (e.g. disliking tags in clothes, or enjoying listening to very loud music), SMD is only considered when sensory-response behaviors impact an individual's ability to engage in and perform daily life tasks [Miller et al., 2007]. For example, a child who is a very picky eater or gags at certain food smells may not be able to engage in family meal time activities and may secondarily have nutritional complications. Similarly, a child who seeks to make loud noises or seeks out intense movement experiences on a regular basis may have problems socially or performing in academic settings. In fact, children with higher rates of atypical sensory behaviors have been shown to have greater incidence of academic difficulties, social-emotional problems, and delays in self-care skills [Ashburner, Ziviani, and Rodger, 2008; Ben-Sasson, Carter, and Briggs-Gowen, 2009; Jasmin et al., 2009].

While responses to sensory stimuli have been most widely studied in pre-school and school-aged children, there is evidence that atypical sensory behaviors may be observed in younger children and may even be an early diagnostic indicator for certain pediatric conditions. Using retrospective video analysis, Baranek [1999] found that infants with autism could be identified on the basis of sensory-motor features at 9–12 months of age. Similarly, children with fragile X syndrome showed a pattern of sensory-motor features (e.g., repetitive leg movements, posturing, less sophistication/repetitive use of objects) at 9–12 months of age that were distinguishable from patterns seen in the autism population, suggesting that sensory-motor features may be important to consider for early differential diagnosis. While motor features of Barth syndrome such as muscle weakness and early gross motor delays have been described, sensory responses may be another important feature to consider in Barth syndrome. Moreover, sensory responses may serve as a potential indicator for early diagnosis and as a feature that may impact functioning and quality of life in this population.

Barth syndrome: clinical phenotype

Since first identified by Barth and colleagues [1983], significant advances have been made in our understanding of the genetic origins of the disorder, as well as subsequent cellular and systems-level alterations. It is now known that Barth syndrome is caused by mutations of the *TAZ* gene at Xq28, which encodes tafazzin proteins [Steward et al., 2010; Spencer et al., 2006]. These proteins, which are deficient in individuals with Barth syndrome, are believed to play an important role in lipid metabolism (as an acyltransferase) and an essential role in the synthesis of cardiolipin, a phospholipid critical for the mitochondrial function enabling conversion of glucose to ATP in neutrophils and other cell types [van Raam and Kuijpers, 2009].

The genetic aberration described above results in the two core clinical manifestations of Barth syndrome, which include cardiomyopathy and neutropenia. Current reports suggest, however, that there is a fair degree of heterogeneity in terms of clinical presentation, with patients presenting with either cardiomyopathy or neutropenia or both concurrently [van Raam and Kuijpers, 2009]. Skeletal myopathy, gross motor delays, and growth retardation are additional common symptoms that also lack heterogeneity in clinical presentation. Spencer and colleagues [2006] reported a high prevalence of growth delay in children with Barth syndrome, though by late adolescence most children had reached the 50th centile for height, indicating some catch-up growth following puberty.

Beyond these core clinical features, few researchers have attempted to describe or quantify associated behavioral patterns or characteristics unique to this population. Hastings and colleagues [2009] described the dysmorphology of Barth syndrome. They indicated that while female carriers of the disorder show no identifiable features; boys with Barth syndrome do show some patterns of facial dysmorphology. These characteristics include having a tall and broad forehead, prominent chin, full cheeks, large ears and deep-set eyes. Though only observed in a small sample of boys with Barth syndrome (N=10), these features are believed to be observable in infancy and persist throughout childhood, with diminished prominence after puberty. Mazzocco and colleagues [2001, 2007] have attempted to identify a cognitive phenotype associated with Barth syndrome. These researchers have found that while boys with this disorder have cognitive abilities (intelligence quotients) and verbal skills in the normal range, they appear to have specific delays in areas of mathematics and visual spatial tasks, particularly in areas of visual motor integration. These authors suggest that while visual motor integration may be associated with the physical phenotype of Barth syndrome (e.g. muscle weakness, fatigue), some aspects of visual perception seem to be deficient as well. These deficiencies were observed in 70% of their sample (n=10) [Mazzocco et al., 2007].

It is currently estimated that around 1/500,000 children each year are born with Barth syndrome [Mazzocco et al., 2007]. Since certain features of the condition such as growth delay and neutropenia are not universally present, early identification of the disorder can be challenging and under-diagnosis is suspected [Hastings et al., 2009; Spencer et al., 2006; Mazzocco et al., 2007]. Similar to fragile X syndrome, cognitive phenotypic patterns and dysmorphic features present in children with Barth are likely not going to be useful in early diagnosis or particularly amenable to early intervention in the first few years of life. The purpose of this paper was to examine the prevalence of atypical sensory processing in children with Barth syndrome, and to explore if phenotypic patterns of sensory responsiveness may be useful in early differential diagnosis and therapeutic intervention.

METHODS

To achieve our study aims, a cross-sectional mixed methods approach was used. The sponsoring university's internal review board approved all processes and consent and assent procedures were completed for all participants.

Sample

Males between the ages of 4–17, diagnosed with Barth syndrome, were recruited through the Barth Syndrome Foundation. Originally 25 children were referred to the study; however, four were excluded due to co-morbid diagnostic conditions (e.g. autism, neurological insult) or incomplete forms. The final sample included 21 boys with a mean age of 9.5 years.

Data collection

For children 4–10 years of age ($n=12$), parents completed a Short Sensory Profile [Dunn, 1999], a 38 item standardized questionnaire that asks about a child's typical responses to a variety of sensory stimuli (e.g. shows distress during grooming). Children and adolescents who were 11–17 years of age ($n=9$) completed an Adult/Adolescent Sensory Profile [Brown and Dunn, 2002], which assesses similar domains of responsivity but was normed on a sample of individuals ages 11–97 years. On both versions of the Sensory Profile, individuals are asked to rank the frequency of sensory behaviors on a five-point scale ranging from *Always* (this behavior occurs 100% of the time) to *Never* (this behavior never occurs). Cut-point scores for the Sensory Profile questionnaires were used to develop three categories of scores based on standard deviation (SD) of the normative sample: Typical Performance (at or above 1 SD below the mean), Probable Difference (between 1 and 2 SD below the mean), and Definite Difference (more than 2SD below the mean).

Qualitative interviews were conducted with children and families over a period of two days at an International Barth Syndrome Foundation conference. All interviews were audio-recorded using standard digital recorders. Families were instructed to complete the Sensory Profile questionnaires prior to the interviews; however, many completed the questionnaires during the interview process while reviewing questions with the researchers. During the interviews, the questions on the Sensory Profile were used to guide questions, and families were asked to elaborate on specific items. For example, if a parent marked *Frequently* for the item "avoids going barefoot", they were probed with questions such as "what textures does he avoid walking on barefoot", "how long have you noticed this behavior" and "does he mind touching those textures with his hands?" When adolescents self-reported for items, they were probed in a similar way with questions such as "tell me more about what you like to eat" or "have you always felt that way about crowds?" When possible, parents of adolescents were asked about the developmental trajectories of specific sensory behaviors, particularly related to feeding, sleeping, and self-care. For example, a parent of a teenager might have been asked, "did he have any problems with feeding when he was younger?" A total of nine interviews were completed and used to validate and expand upon questionnaire data.

Data analysis

Sensory Profile questionnaires were scored [Dunn, 1999; Brown and Dunn, 2002] and entered into a statistical analysis software program (PSAW 18.0). Descriptive data analysis was run separately for subjects completing the Short Sensory Profile and the Adult/Adolescent version since the domains for each are slightly different.

Audio recordings of the interviews were transcribed and all identifiable information (i.e. names, place of residence) was removed. Following transcription, all text files were

imported into a qualitative software analysis program. Once imported, each author read each interview through two times. Based on the initial readings a preliminary code sheet was developed. Codes were further refined with the third and fourth reviews of the transcripts. Codes reflected primary sensory responsivity patterns and secondary features related to child development, intensity of sensory behaviors, and impact on function. Further analysis from several subsequent reviews of the data resulted in the collapse and expansion of codes, the development of categories, and the emergence of themes. The final code sheet is presented in Table I.

RESULTS

Short Sensory Profile

Total scores on the Short Sensory Profile indicated that 75% of the children in our sample who were between 4–10 years of age fell into either the Probable Difference or Definite Difference range with regards to their overall sensory processing abilities. All of the children in this sample (n=12) had scores in the Definite Difference range for the sub-test of Low Energy/Weak. At least half of the sample also had scores in the Probable Difference or Definite Difference range in the domains of Taste/Smell Sensitivity, Auditory Filtering, and Under-responsive/Sensation Seeking (Table II).

Adult/Adolescent Sensory Profile

In our sample, children that were 11–17 years of age who completed the Adult/Adolescent Sensory Profile performed relatively similar to the normative sample on all four quadrants related to sensory processing. Sensation Seeking behavior was seen less often in older children with Barth syndrome compared to the normative sample (Table II). While the scoring on this version of the Sensory Profile does not allow for analysis of specific sensory domains, we did visually examine each of the nine forms for frequently reported items. In the area of taste/smell processing, six out of the nine subjects reported that they either found most food to be bland or added spice to their food frequently or always. Four out of the nine subjects reported that they only ate familiar foods. One third of the sample (n=3) also reported being frequently or always bothered by strong odors. In the area of auditory processing, three of the nine respondents either reported having difficulty concentrating when there was competing noise in the background or that they startled easily when there was a loud or unexpected noise. An additional two subjects reported that they frequently limited distractions when working.

Family and child interviews

Based upon our analysis of the family and child interviews, three primary themes emerged related to sensory processing in Barth syndrome. Feeding and Eating sensory issues referred primarily to taste, textures, and scent. Sensory Intensity described intensity of responses to movement, touch and food experiences. Sensory Impact elucidated sensory consequences for the child and family.

Sensory issues related to feeding and eating

Eight of our nine sources indicated responses in one of the following categories: taste/smell seeking (7/9), taste smell sensitivity (4/9), strong texture preferences or tactile food sensitivity (5/9), or a history of gagging in response to food or food smells (4/9). In the area of taste/smell seeking it was noted that many of the children had strong preferences for salty and cheesy foods with one child directly stating “I love salt.” One parent expressed concern about her son’s eating habits noting, “He doesn’t eat and it’s hard to tell if it’s a sensory problem or a Barth problem; but he only eats like salty-cheesy foods.” Specific food

preferences identified by parent and child respondents included grilled cheese sandwiches, vinegar, garlic, French fries, salt and vinegar potato chips, nachos with cheese, and macaroni and cheese. One child explained the preparation of his favorite snack: “I’ll take vinegar, put salt in it, take ketchup put it in the vinegar, put (more) salt and dip my fries in it.”

Despite strong preferences for certain foods or flavors, many children with Barth were also identified as being picky eaters with either strong taste, smell or texture aversions. In the area of food textures, one mother noted “It’s more what he doesn’t like; he doesn’t like mushy. He doesn’t like mousse or whipped cream. He will immediately gag and not eat it... Even regular food that he likes, if he chews on it too long and it gets mushy he starts gagging.” Other children were reported to have strong aversions to lumpy foods such as soup or smooth/creamy foods such as yogurt. Another parent noted, “He thinks he wants yogurt, he likes the idea of it, but he won’t eat it...or smoothies; he thinks he wants a smoothie but he won’t eat it.” Interestingly, food preferences did not seem to be strongly related to the boys neuromuscular weakness; with only one family noting that steak was difficult for their child to chew and therefore he didn’t often choose to eat it.

Problems with gagging were brought up by four respondents. While gagging can be elicited through difficulting coordinating food in the mouth, this did not appear to be the case in our sample. Both taste and smells were noted to elicit the gag reflex with one mother remarking “...he used to be real bad about gagging; but they weren’t necessarily (all) food smells”. In addition, three parents noted that the problems with gagging were more noticeable when the child was younger.

Intensity of sensory behaviors in Barth syndrome

While we saw different types of sensory behaviors (both over and under-responding/seeking) across multiple sensory domains (e.g. tactile, auditory), what emerged from our interviews was that many of these behaviors were significantly more intense than what tends to be reported in the typical (non-diagnostic) population. Often this intensity was identified by a strong emphasis on the words *always* or *never* when parents or children described engagement in specific tasks. For example, when asked about their child’s willingness to go barefoot two families responded with opposite extremes of preference for their child. One family stated “He *loves* barefoot! He *prefers* barefoot”; while another family noted “Sand *never*, he *never* wants to go barefoot in the sand. He’s not *so* bad with grass”.

Some of the intense behaviors reported were again related to issues of feeding and eating. One family noted intense sensory experiences their child had related to temperature: “And he freaks out about hot as well. Like if we just heat something up, it immediately has to go in the fridge or we have to add ice to it. So he wants it warm, but if it’s anything more than luke warm...” Other children and families emphasized specific preferences like “he *loves* things spicy” or “he’s a *very* picky eater”.

Finally, some comments related to movement and touch also emerged as being particularly intense or salient for children with Barth syndrome. Unsurprisingly for this diagnostic group, there were several reports of intense levels of muscle weakness or fatigue noted. One family compared their child’s functional grasp in relation to non-Barth peers: “Yes, it’s hard to put it into a degree. It’s weaker than typical but he does color, he does write... (but) it’s *always* weaker than the average child”. Muscle weakness and fatigue may also relate to one child’s reported extreme sensitivity of heights. This family stated “Like if he gets sat on anything higher than a normal seat he wants someone there with him. He’s immediately fearful he’s going to fall. He’ll start screaming ‘I’m going to fall, I’m going to fall’”. Touch sensitivities were less common but intense (non-eating related) tactile behaviors were noted in 3/9 families. One family reported that their child was so bothered by getting his hands

messy that he refused to do painting at school. Another described distress during grooming activities: "...like when I'm washing his face or clipping his nails it's the pulling away and the whining, 'no, no, don't'... And the anticipation of cutting the nails, he doesn't like the way that feels".

Sensory behaviors influence routines and impact child and family function

Auditory processing issues were associated with participation in school and extracurricular activities for children with Barth. One parent reported that their son was moved out of a regular classroom because he couldn't tune out the noise in that environment. Another family reported that, as a compensatory strategy, their child would leave the room when there was a lot of noise or if he was having difficulty paying attention. When asked specifically about responses to sound in the environment, one parent noted "he isn't good at birthday parties or things like that".

Food issues also came up in relation to impact on family function and changes in daily routines. Families reported buying certain preferred foods or altering foods (e.g. by adding salt or spice) to meet the specific preferences of their child with Barth syndrome. Another family expressed more severe concerns about their child's overall health due to dietary restrictions: "My concern is always with his growth and how to always measure that. Because he does stick to chicken nuggets, chips, and spaghetti O's and chicken noodle soup. He's very consistent in what he wants; hardly ever does he step out".

Lastly, motor issues of weakness and fatigue were described in relation to family function or need for compensatory strategies by three of the nine interviewed families. One family noted that due to their son's fatigue they would often do a lot of their play on the floor instead of standing or sitting upright at the table. One child self-reported that when his hand got tired from writing his strategy was simply to wait until math, a subject that required less handwriting. Another family noted that when their child became fatigued he would get very fidgety, "like he's trying to keep himself awake".

DISCUSSION

The purpose of this study was to describe sensory processing issues in children with Barth syndrome and determine if there were phenotypic atypical sensory processing behaviors specific to this population. In children 4–10 years of age, total scores on the Short Sensory Profile indicated that 75% of our sample had scores suggestive of atypical sensory processing. This total score, however, appeared to be largely driven by scores on the sub-test of Low Energy/Weak, a domain in which 100% of our sample had scores in the Definite Difference range. In sensory integration theory, symptoms such as low muscle tone and poor postural stability are associated with functioning of the vestibular and proprioceptive sensory systems; the category of Low Energy/Weak, therefore, was designed to capture these challenges in children who may have disorders of these areas of sensory processing. While we cannot rule out vestibular or proprioceptive deficits in children with Barth, we can assume that the uniform presence of atypical scores in this category are driven not by sensory processing deficits but by the metabolic fatigue and weakness brought on by their genetic condition.

More interesting and relevant to this study were the atypical sensory behaviors reported in other areas of sensory processing. The most consistent finding was that children with Barth syndrome appear to have atypical food-related sensory behaviors that seem to stem from taste, smell or texture sensitivity or a need to seek out specific taste sensations. This finding was noted in both our older and younger group of children with Barth and was validated by the child and family interviews. By triangulating this data we found that most children in our

sample were restricted in what types or textures of food they would consume yet had strong preferences for intensely salty, spicy and/or cheesy foods. Interestingly, these specific feeding behaviors were reported by some parents to have been present from a very early age, often marked by children having strong gag reflex responses to certain tastes and smells. In some cases these taste/smell sensitivities became less pronounced over time; in other children it appeared that compensatory strategies were put into place so that the child's diet was adjusted to meet their specific sensory preferences. While most families were comfortable with making dietary changes based on their child's sensory preferences, some families did express resultant worries regarding overall nutrition and related growth. To our knowledge, no studies to date have examined the effects of sensory-based dietary changes on health status in boys with Barth syndrome.

Auditory processing concerns were also identified in both our younger and older sample of children with Barth and supported by our qualitative data analysis. While not as prominent as feeding issues, multiple children in our sample reported having difficulty with filtering out sounds in the environment or being bothered by loud sounds. These auditory processing difficulties were reported to influence participation in academic tasks and choice of participation in extracurricular activities (e.g. not going to birthday parties or to the mall with friends). Unlike feeding issues, however, these auditory processing difficulties were not described by parents as being problematic during early childhood (before the age of 3) and appeared to become more evident as the demands of school or social expectations (e.g. hanging out with a large group of friends) increased. It is possible, therefore, that auditory processing difficulties may be present in younger children with Barth, but that functional implications are not evident until closer to school age where a child has to engage in more complex and challenging environments.

While the purpose of this study was to describe and quantify sensory processing issues in boys with Barth, we can begin to speculate about causes for these behaviors in this population. In other genetic disorders, like Smith-Lemli-Opitz syndrome, dysregulation in cholesterol metabolism results in differences in oral, tactile, visual and auditory processing (sensory hyperreactivity/hyper-sensitivity) [Eapen, 2007; Tierney et al., 2001]; possibly there is a direct connection between lipid metabolism and sensory sensitivity or sensory cravings in children with Barth? Another possibility is that boys with Barth syndrome have a sodium deficiency, common in conditions such as hypovolemia, and that sodium depletion may be the root cause of taste disturbances and salt cravings [Geerling and Loewy, 2007]. To our knowledge, a thorough examination of nutritional status and/or blood volume in children with Barth has not been conducted. Future studies may also investigate the relationship between disruptions in mitochondrial function and sensory issues in children with Barth syndrome. Some authors have proposed mitochondrial dysfunction as a possible cellular basis for symptoms of autism spectrum disorders [Giulivi et al., 2010], a condition in which 70–90% of affected children have some type of sensory processing disorder, including taste/smell sensitivity and problems with auditory filtering [Baranek et al., 2006; Lane et al., 2010]. While it is clear that children with Barth syndrome are not autistic, there may be metabolic and/or cellular mechanisms underlying sensory-based behaviors within these disorders which warrant future study.

CONCLUSION

This study was the first to examine sensory response patterns in children with Barth syndrome. Based upon our preliminary findings it appears that sensory issues related to feeding and eating are important to consider for boys of all ages with Barth syndrome. While challenges with auditory filtering and auditory sensory sensitivity can impact school and family functioning in older children, auditory issues can also be screened for at a very

young age. Early identification of these observed sensory issues can potentially assist in differential diagnosis. Early identification can also create opportunities for early interventions that may minimize detrimental functional impacts of the atypical sensory responses described here. Future studies should examine a larger sample of children with Barth syndrome and include children under the age of three diagnosed with Barth or at risk for having Barth syndrome (i.e. born to a known carrier). Sensory responses should also be examined over time while investigating functional impact and coping strategies used by children and families. Sensory response patterns may serve as a clinical phenotype potentially useful in guiding both diagnosis and intervention.

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Table I**Code Sheet for Sensory Interviews**

Primary Codes	Sources	References	Sample Quotes
A. Sensory Sensitivity			
a. Tactile	7	18	<i>I've got to take all the tags off his clothes; his shoes have to be exactly right, his socks have to be exactly right</i>
b. Taste/Smell	4	7	<i>I don't like bad smells. I have a very sensitive smell and sensitive taste</i>
c. Movement	2	3	<i>Like if he gets sat on anything higher than a normal seat he wants someone there with him. He's immediately fearful he's going to fall.</i>
d. Auditory	2	5	<i>(when sounds are loud or unexpected) Hands up on ears then he'll cry</i>
e. Visual	2	2	<i>He always goes "the sun's in my eyes, the lights in my eyes"</i>
B. Under-responsive/Sensation Seeking			
a. Tactile	6	10	<i>He loves going barefoot; he prefers going barefoot.</i>
b. Taste/Smell	7	9	<i>I love salt!</i>
c. Movement	3	3	<i>He loves being upside down, I think more than he likes being right side up</i>
d. Auditory	4	5	<i>Oh he loves strange noises and always screams just to make noise</i>
e. Visual			N/A
C. Low Energy	7	14	<i>He still can't sit up on his own</i>
D. Auditory Filtering	2	2	<i>They moved him out of a regular classroom because he can't tune it (the noise) out</i>
Secondary Codes	Sources	References	Sample Quotes
1. Development	4	10	<i>From birth he's always had a good grasp</i>
2. Intensity of Behavior	7	30	<i>Always hugging and touching; even strangers!</i>
3. Compensatory Strategies	6	11	<i>(If there is a lot of noise in the room) He'll leave the room</i>
4. Therapies	2	3	<i>... we had to do a lot of play therapy in sand, play dough...we did brushing.</i>
5. Reference to Gag Reflex	4	4	<i>...he couldn't put anything in his mouth without gagging</i>
6. Impact on Function	4	9	<i>He isn't good at birthday parties or things like that</i>

Note: Sources= number of participants, References= number of times code was referred to in the interview

Table II

Short Sensory Profile Scores for Boys with Barth (n=12)

Domain	Mean	Std. Deviation	Typical Performance	Probable Difference	Definite Difference
Tactile Sensitivity	30.3	5.6	75%	-	25%
Taste/Smell Sensitivity	13.1	6.6	50%	16.7%	33.3%
Movement Sensitivity	12.6	3.4	58.3%	25%	16.7%
Under-responsive/Seeks Sensation	26.9	5.1	50%	25%	25%
Auditory Filtering	22.4	6.2	50%	16.7%	33.3%
Low Energy/Weak	12.3	4.6	-	-	100%
Visual/Auditory Sensitivity	19.6	4.8	75%	8.3%	16.7%
Total Score	137.1	24.0	25%	25%	50%

Table III

Adult/Adolescent Sensory Profile Scores for Boys with Barth (n=9)

Domain	Mean	Std. Deviation	Less Than Most People	Similar to Most People	More than Most People
Low Registration	32.0	6.7	22.2%	66.7%	11.1%
Sensation Seeking	42.1	5.9	44.4%	55.6%	-
Sensory Sensitivity	48.0	8.7	11.1%	77.8%	11.1%
Sensation Avoiding	42	6.9	33.3%	55.6%	11.1%