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## Challenging Behavior in CHARGE Syndrome

Key Words: CHARGE, behavior, deaf-blind, psychiatric, developmental disabilities, OCD, autism, tics, ADHD

### Abstract

Little is known, beyond anecdotal reports, concerning the challenging behaviors of some children with CHARGE Syndrome. One hundred respondents from the US (74%) and 7 foreign countries, primarily mothers (91%), completed a web-based survey regarding the behaviors of a person with CHARGE (median age of 7). Included was a medical history and a list of 71 behaviors based on the diagnostic categories most frequently reported anecdotally. Findings supported these reports. Behaviors typical of autistic disorder, attention deficit/hyperactivity disorder, obsessive-compulsive disorder, Tourette syndrome, and deaf-blindness were characteristic of these children. Those who were deaf-blind received higher ratings on these challenging behaviors.

## Introduction

CHARGE Syndrome, or Association, was first identified by Hall (6) in 1979. The acronym was suggested by Pagon et al (16), based on common features: C – coloboma of the eye (missing part of iris and/or retina); H – heart defects; A – atresia of the choanae (bony or membranous blocking of nasal passage); R – retardation of growth and/or development; G – genitourinary anomalies; E – ear anomalies and/or deafness. While all six features may appear, there is wide variation in both occurrence and severity. This has led to a more refined diagnostic system by Blake et al (2) with four major features and a number of minor. The prevalence of CHARGE is generally estimated as 1:10,000 (10).

Complications with CHARGE may include multisensory impairment including deaf-blindness. A result of the multisensory impairment may be poor communication skills. Another complication is impaired ability to swallow food, and many children are tube fed by gastrostomy. Children with CHARGE very frequently have damaged or missing semi-circular canals in their ears resulting in impaired balance and ocular coordination. Children with CHARGE are often short statured and may have mild to severe developmental delays.

Behavior problems, often described as “autistic-like”, are another complication. However, there has been little investigation of these. Denno and Bernstein (4) conducted the first study of behavior in CHARGE. They compared seven students with CHARGE at Perkins School for the Blind with a control group matched for age, sex, and functional level. The control group included five with Rubella syndrome, one with infantile encephalitis, and one with multiple disorders due to prematurity. The main identified difference between the two groups was compulsive behaviors, where the students with

CHARGE displayed more compulsive behaviors and displayed more negative behaviors when staff redirected or interrupted their compulsive behavior.

A second report came from Sweden by Fernell et al (5). These authors reported on three children, all of whom had elevated scores on the Autism Diagnostic Interview. They suggested that a neuroendocrine dysfunction in CHARGE might contribute to the autistic behavior.

As Lewis and Lowther (14) note, by school age most medical problems for children with CHARGE are well managed, and concerns become more focused on learning and behavior. However, they point out, "There has been very little investigation into the behavioural aspects of CA, and this is an area that deserves careful research in the future" (p. 72).

Based on his contacts with parents, Hartshorne (8) addressed some of the behavioral diagnoses children with CHARGE received: autistic disorder, attention deficit/hyperactivity disorder, obsessive-compulsive disorder, and tic disorder. He maintained that many of these behaviors could be attributed to multisensory impairments, such as deaf-blindness.

Information on behavioral difficulties in CHARGE is still largely anecdotal. While specific clinical studies are needed, so too is information regarding the prevalence and nature of the behavioral difficulties. The purpose of this study was to investigate the kinds of behaviors seen in children with CHARGE in a larger sample than has previously been studied. A secondary purpose was to look for any associations that might suggest etiological factors. Specifically, these were hospitalizations, number of medical conditions, vestibular functioning, and deaf-blindness.

## Methods

### *Ethics*

Procedures for this study were approved by the University's Institutional Review Board.

### *Sample and Procedures*

In 1998 a listserv for parents and others interested in CHARGE Syndrome was established (<http://groups.yahoo.com/group/CHARGE/>). This list now has over 400 members, although not all of them are active and not all of them are parents. It was decided, with permission from the list manager, to solicit respondents from the list.

In order to facilitate the gathering and recording of data, a web-based questionnaire was developed, using Microsoft FrontPage. An email message was sent to the list, giving the web address for the questionnaire, and asking parents to log in and complete the form. Upon pressing "submit" the data went to a Microsoft Access file, which was later converted to SPSS for the analyses.

In addition to the listserv, interested individuals on the listserv sent invitations to other parents. For example, it was distributed to a separate list in Canada, and an invitation to participate appeared in the Australasian CHARGE Association newsletter.

### *Questionnaire*

The questionnaire began with some basic demographic information in order to characterize the respondents, and the gender and age of each child with CHARGE. The second part of the questionnaire gathered information about medical conditions and hospitalizations of the child. This portion was based on a form developed by N. Hartshorne (7). Questions on hearing and vision impairment were based on the deaf-

blind census form used by DB Central, Michigan Services for Children and Youth Who are Deaf-Blind ([www.dbcentral.org](http://www.dbcentral.org)).

The remainder of the questionnaire consisted of a list of 71 behaviors that respondents scored on a five-point likert scale for their child. Hartshorne (8) noted that the behavioral diagnoses he found to be most frequently given to children with CHARGE were autistic disorder, attention deficit/hyperactivity disorder (ADHD), obsessive-compulsive disorder (OCD), and tic disorder. He maintained that many of these behaviors were common to children who are deaf-blind. We therefore contacted individuals with expertise in all of these areas except for tic disorders, and asked them to provide a list of typical behaviors demonstrated by children with these diagnoses. These lists, plus two questions for verbal and motor tics based on the American Psychiatric Association's *Diagnostic and statistical manual of mental disorders* (4<sup>th</sup> ed.) (*DSM-IV*) (1), comprised the 71 behaviors.

It should be noted that the 71 behaviors are not necessarily discrete. This reflects overlapping behaviors, particularly in the autistic disorder and deaf-blind categories.

## Results

### *Respondents*

One hundred responses were received. For this reason, actual numbers and percentages are the same in the study's results. Ninety-one of the responses were from mothers, four from fathers, and five from others, including one person with CHARGE. Of those with CHARGE, 43 were male and 56 female (one no response). Ages ranged from under one to 30 years, with a median age of seven and a modal age of four. As seen

in Table 1, nearly three quarters were from the US (in 26 states), while the other 26 represented eight foreign countries.

Table 1 presents information regarding medical history. There was considerable variability in when children came home from the hospital after birth, with 20% coming home at the same time as the mother, and 20% coming home after more than 12 weeks. While four never had any surgeries, 29 had one to four surgeries, and 23 had more than 12. Two children had never been hospitalized, 30 had experienced one to four hospitalizations, and 29 had experienced more than 12. The modal time period for being able to walk independently was 18 months to three years of age. However, nearly a third learned to walk between the ages of three and five.

CHARGE may involve a number of different anomalies. The medical problems of these 100 individuals are shown in Table 2. The most frequent were delayed motor milestones, coloboma, and sensorineural hearing loss. In general, the percentages are similar to those reported by Blake, et al. (2).

Table 3 shows the degree of hearing and vision loss for this group. There were more hearing than vision problems. The number who had no impairment in either vision or hearing was small.

Respondents indicated the label of their child on an Individualized Education Program (IEP). These are reported in Table 4. Many children had multiple labels. The most common label was hearing impaired. It is interesting to note that while 16 children were labeled as deaf-blind, another 14 children were labeled as both hearing impaired and visually impaired and so might be appropriately labeled as deaf-blind.

Respondents were also asked if the child had received a diagnosis for their behavior (see Table 4). All of the diagnoses listed were conditions that had been included in the questionnaire, providing some evidence of validity to the anticipated range of diagnoses.

### *Behaviors*

A total of 71 behaviors were listed for respondents to rate on a five-point likert scale, with a “1” representing "very much not like my child" and a “5” "very much like my child". The midpoint (“3”) was “unsure.” Two kinds of analyses are presented. First, the number of individuals who received scores of four or five on each behavior was calculated. Scores of four or five mean the child does demonstrate the behavior. This allows for comparisons between the behaviors. Second, scores on the likert-type scale were totaled for the 71 behaviors overall and within each behavioral category. While this masks some of the variability among those with CHARGE on the behaviors, it allows for the examination of relationships between behavior and other variables.

Table 5 lists the 71 behaviors within the five behavioral categories, ranked by the number of scores above three, which are shown next. The following two columns present the scale mean and standard deviation for each behavior. Overall, the three behaviors with the greatest frequency and highest mean were: (1) Extreme preference for certain toys, people, food, etc. (deafblind); (2) Restricted range of interests and/or pre-occupation with one narrow interest (autistic disorder); and (3) Significant difficulty in ability to make same age friendships (autistic disorder). The final two columns of Table 5 are discussed below.

Fifteen of the 71 behaviors were scored above a three for more than 50% of the persons with CHARGE, five each in the autistic disorder, ADHD, and deaf-blind categories. Using a criterion of least one-third (33%) of those with CHARGE being rated as demonstrating the behavior, then 15 of the 25 autistic disorder, nine of the 10 ADHD, one of the 13 OCD, 15 of the 21 deaf-blind, and one of the two tic behaviors qualify as commonly occurring within this population. This is a total of 41 behaviors out of the 71 (58%).

All 100 persons with CHARGE received some scores above three on the behaviors. Two received only two scores above three, while one received 51. The median was 25, the mean 24.71 and standard deviation 11.89.

Due to different numbers of items for each of the diagnostic categories, raw means could not be compared, and so scale scores were used. ADHD had the highest scale mean (3.0), followed by autistic disorder (2.7), deaf-blind (2.6), and tic (2.5), with OCD (2.0) having the lowest.

*Interrelationship of the behaviors.* Table 6 shows the correlation coefficients for the five categories of behaviors using total scores. As can be seen, all of the categories were significantly correlated with each other, with correlations ranging from a low of .31 between scores of the categories of OCD and Tics, and a high of .66 between autistic disorder and deaf-blind.

*Association between age and problem behaviors.* A Pearson correlation was calculated between age and total behaviors and the behavior categories. Caution needs to be taken in interpreting this and the following correlations, as a large number of correlations were calculated. We have reported the traditional two-tailed probability

results; however, as we were testing predicted relationships, one-tailed probability estimates are appropriate, and so each probability may be divided in half. As shown in Table 7, small but significant correlations were found for total behaviors, autistic disorder, OCD, and ADHD. As children with CHARGE age, they are more likely to develop problem behaviors in these three categories.

*Potential explanations for problem behaviors*

*Hospital Experience.* It has been suggested that the experience of children with CHARGE in the hospital contributes to their later development of difficult behaviors. Thus correlations were calculated between the various behavior categories and number of hospitalizations, number of surgeries, and length of time in the hospital after birth. While these three were intercorrelated, none of them was significantly associated with total behavior or the various behavior categories (see Table 7).

*Number of Medical Conditions.* It has been suggested that the number of medical conditions a child with CHARGE has may be associated with the development of problem behaviors. For that reason a total score was derived for the conditions listed in Table 2, and this total score was correlated with behaviors. Significant, but small, correlations were found with autistic disorder (.224) and tics (.214) (see Table 7).

*Age of Walking.* Age of walking is likely related to vestibular difficulties as well as degree of deaf-blindness. Correlations were calculated between age of walking and the behavior categories. Statistical significance was found for total behaviors (.20) and deaf-blind behaviors (.26), although once more these were small (see Table 7).

*Deaf-blindness.* Using the responses to the questions on hearing and vision, a category of deaf-blindness was created for those children whose hearing and vision were

both affected. To qualify, the child had to have at least a mild hearing loss in one ear, together with at least low vision in one eye. Based on this, 66 of the children were categorized as deaf-blind and 30 as not deaf-blind (4 cases were missing). Some of these cases might not meet national, state, or local criteria as deaf-blind, but they all do have both a visual and a hearing impairment in at least one ear and eye.

These 66 cases were examined for their correspondence with those children who were identified as having an IEP label of either deaf-blind or both vision and hearing impaired. All but one of the 30 cases that had either a deaf-blind label or both hearing and vision impairment labels were among the 66 children so categorized. The one case that was not so categorized did indicate a label of deaf-blind, but also indicated no vision impairment.

In order to determine whether children who were deaf-blind were more likely to have behavior problems than those who were not, the chi square statistic was calculated for each of the 71 behaviors comparing the 66 deafblind cases with the 30 not deafblind. They were compared on the basis of demonstrating the behavior (scale score above 3) or not clearly demonstrating the behavior (scale score of three or below). The 23 analyses that achieved statistical significance are shown in Table 5. Nearly half of these were in the autistic disorder category. Being deafblind does seem to be related to more problem behaviors.

In addition, independent t-tests on the total scores for each of the behavior categories were conducted. The results are in Table 8. As can be seen, children who were deaf-blind were more likely to have higher scores on the total behaviors, and on autistic disorder, deaf-blind, and tic.

## Discussion

This study's primary purpose was to identify the typical behaviors of children with CHARGE in a large sample. This study took the approach of listing behaviors that are typical of psychiatric diagnostic conditions often associated with children who have CHARGE. Support for this approach was found in the list of behavioral diagnoses given by the respondents. It is also supported by the degree of intercorrelation among the diagnostic categories.

Children with CHARGE are quite variable in their medical characteristics, and this study suggests there is considerable variability with their behavior as well. The three behaviors from the list that were most frequently scored as present occurred in 61 to 69% of those with CHARGE, meaning that even for the most frequent behavior, 31% did not demonstrate it. The range of scores for the 71 behaviors was fairly large as was the standard deviation. On average, respondents were somewhat unsure that each behavior reflected their child, although on average each individual received scores of four or five on 25 of the 71 behaviors.

This study provides support for the behavioral diagnostic categories being applied to children with CHARGE. While answers to these questions cannot provide a diagnosis, the 71 behaviors listed are, in general, fairly disturbing behaviors. It is difficult to point to one diagnostic category as the most severe, as there were different numbers of questions in each, and some overlap in behaviors. What is perhaps more important to note is the high degree of intercorrelation among the five categories. Thus, rather than claiming that behavior in CHARGE is more autistic than compulsive, for example, one

might rather say that behaviors in all five categories may be characteristic of CHARGE to some degree.

The top 15 behaviors, those where more than 50% of persons with CHARGE received scores above three, are of interest. These were evenly distributed between the categories of autism, ADHD, and deaf-blind. Some of the behaviors overlap, for example, fascination with shiny objects or things that reflect light (autism) is similar to stares at light sources (deaf-blind), and restricted range of interests (autism) is similar to extreme preference for certain toys, etc. (deaf-blind). One wonders how much the high activity level reflected in the ADHD behaviors is associated with stereotyped body movements (autism) and vocal self-stimulation (deaf-blind). There were not enough subjects for exploratory factor analysis, however this might be a focus for future research.

While many parents report, and Denno and Bernstein (4) found, that OCD behaviors are very characteristic of their children, there were no OCD behaviors in the top 15, and only one above the 33% criterion. However, the literature on OCD in children is not as extensive as that for adults (15) and it is possible that the kinds of OCD behaviors most frequently seen by parents of children with CHARGE were not listed in this study. Further research is needed in this area.

Motor tics were more frequent than verbal tics. This could be consistent with the activity noted in ADHD and stereotyped movements. Twenty-two children were rated as high on both motor and verbal tics, consistent with a possible diagnosis of Tourette syndrome.

Multi-sensory impairment, specifically deaf-blindness, was found to be related to behavior in this study. According to Kates, Schein, and Wolf (12), autistic-like behaviors

commonly accompany deaf-blindness. They list behaviors such as withdrawal, lack of affect, an inability or disinclination to relate to others, self-abuse, self-stimulation, and perseveration, as common among persons who are deaf-blind, and all of which were included as behaviors in our autistic disorder category. Other behaviors in our autistic disorder category that have been associated with deaf-blindness include light gazing, spinning and rocking (13), stereotypy (17), and social skills deficits (18). There is less literature on ADHD, OCD, or tic behaviors in deaf-blindness, but given the results of this study, that may be worth examining. Hindley (11) has discussed the prevalence of attention deficit disorder, affective disorders, and autistic disorder in cases of hearing impairment.

This study did find an association between age and total number of behavior difficulties, particularly those associated with autistic disorder and OCD. Age at which the child walked was also associated with total number of behaviors, and deaf-blindness. Given the number of correlations calculated, this finding should be considered tentative. However, to the extent that deaf-blind behaviors indicate deaf-blindness, this makes sense. The association with age at walking could also suggest a possible association between behavior difficulties and vestibular functioning, as balance problems are associated with difficulties in learning to walk. However, more research is needed to address this relationship.

It has been suggested by parents that the behaviors could be associated with negative medical experiences, such as number of surgeries. This was not supported in our study, as number of surgeries, hospitalizations, and length of stay in the hospital after birth were not significantly associated with behaviors. The number of medical conditions

was also not associated with the total behaviors, but it was with autistic disorder behaviors and tics. This could suggest that medical involvement creates more stress leading to social withdrawal and tics.

### *Implications*

The behavioral difficulties in CHARGE need to be addressed by medical and particularly educational professionals. Parents and teachers need support in coping with these behaviors. This study did not address diagnosis directly, but rather the presence or absence of typical behavior associated with diagnoses. It is an open question as to whether children with CHARGE should receive psychiatric diagnoses. Because these behaviors are so common in CHARGE, it may not make sense to add diagnoses on to that of CHARGE, such as CHARGE and autistic disorder, or CHARGE and ADHD, but rather to recognize that a diagnosis of CHARGE may indicate the potential development of behaviors that are like those in other disorders. However, it can also be argued that the psychiatric diagnoses help to describe the problem behaviors and may result in more appropriate treatment. Certainly an awareness of the prevalence of these behaviors in CHARGE should lead to the implementation of preventative behavioral interventions. Children with severe disabilities may be disciplined differently by parents, and so develop negative behaviors. Parent training and the presence of a behavioral consultant early in the child's life are recommended.

Pharmacological intervention is becoming common for children with CHARGE, based on anecdotal reports from parents on the CHARGE listserv and at conferences. The use of medications in children with CHARGE requires extremely close monitoring, due to the associated medical conditions and the report of parents that many drugs have

unusual and sometimes paradoxical effects on their children. It may be wise to recognize that while a child with CHARGE may show, for example, OCD behaviors, these behaviors may be due to unique causes such as sensory impairments so that the drugs that may generally be effective for OCD might not be effective with CHARGE. However, recognition that these behavioral difficulties are common in CHARGE should alert the medical and psychological professions to the possible need for pharmacological intervention when assessment indicates the presence of concurrent behavioral conditions.

There is increasing understanding of the importance that all behavior is used to communicate something to other people (3, 9). The key to behavioral intervention from this perspective is to increase communication skills and to help the child find alternative means of communication. A temper tantrum may be effective in helping a child to get what they want, but there are more appropriate communication methods. In a similar vein, autistic-like, ADHD, OCD, and tic behaviors may be communicating stress, over or under arousal, pain, confusion, or general unhappiness. Intervention should take these and other possible functions of the behavior into account (3).

Future research should refine the typical kinds of behavior found in CHARGE. It also should compare and contrast children with CHARGE who have behavioral difficulties with those who do not, in an effort to identify possible causal agents. Finally, future research should be directed toward sorting out behaviors that are due to the neurological/genetic/medical basis of CHARGE and those that are a response to the multi-sensory impairments and social experiences, in the interest of better understanding how the challenging behaviors can be avoided or, when present, treated.

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Table 1

*Child Characteristics*

	N & %
Country	
US	74
Canada	7
Australia	6
New Zealand	5
Germany	3
Austria	1
England	1
Scotland	1
South Africa	1
Age Home from Hospital	
With Mom	20
< 2 weeks	10
2-6 weeks	27
7-12 weeks	23
> 12 weeks	20
Number of surgeries	
None	4
1-4	29
5-8	20
9-12	21

>12	23
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#### Number of Hospitalizations

None	2
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1-4	30
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5-8	28
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9-12	11
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> 12	29
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#### Age Walked

< 18 months	5
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18mo – 3 yrs	40
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3 – 5 yrs	31
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6 – 7 yrs	4
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> 7 years	2
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Table 2

*Medical Problems in the Sample*


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Condition	Number and Percent (N=100)
Delayed Motor Milestones	93
Coloboma	86
Sensorineural Hearing Loss	80
Heart Defect	75
Swallowing Problems	74
Frequent Middle Ear Infections	71
Vestibular Problems	63
Growth Deficiency	62
Choanal Atresia or Stenosis	54
Facial Palsy	48
Genital Hypoplasia	39
Cleft Lip or Palate	27
Spine Anomalies	27
Renal Problems	23
Hypoxia	18
Tracheosophageal Fistula	14
Hand Anomalies	13
Abdominal Defects	13

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Table 3

*Degree of Hearing and Vision Loss*

Hearing	N and %	Vision	N and %
None	8	None	25
Mild	15	Low	25
Moderate	11	Legally blind	26
Moderate-severe	19	Only light perception	3
Severe	16	Totally blind	2
Profound	24	Cortical impairment NOS	1
Progressive loss NOS	1	Progressive Loss NOS	9
Further testing needed	5	Further testing needed	6

Table 4

*Individualized Education Program (IEP) Label and Behavioral Diagnoses*

IEP Label	N
Hearing	43
Multiply/Severely multiply impaired	21
Vision	17
Deaf-blind	16
Developmentally delayed	13
Physical or other health impairment	11
Speech/Language	4
Learning disabled	1
Behavioral Diagnosis	N
Attention Deficit with or without hyperactivity	7
Autistic disorder or autistic-like	6
Pervasive Developmental Disorder	6
Obsessive Compulsive Disorder	3
Tourette Syndrome	2

Table 5

*List of Behaviors in Categories, Scores on the Behaviors, Comparison of Those Who Are and Are Not Deaf-Blind*

	Scores >3	Mean	SD	Chi * sq	p
<b>AUTISTIC DISORDER</b>					
Restricted range of interests and/or pre-occupation with one narrow interest	66	3.75	1.47	5.46	0.020
Significant difficulty in ability to make same age friendships	61	3.63	1.53	4.03	0.045
Preference for solitary play	55	3.30	1.57	8.44	0.004
Stereotyped (repetitive) body movements	54	3.10	1.64		
Fascination with shiny objects or things that reflect light	52	3.23	1.63	4.75	0.029
No or abnormal social play	47	2.94	1.61	4.26	0.039
Tends to interpret information literally	46	3.39	1.17		
Looks out of peripheral vision	46	3.08	1.54	7.90	0.005
Distress over changes in trivial or unimportant aspects of the environment	45	2.89	1.60		
Unreasonable insistence on following routines in precise detail	42	2.93	1.39		
Seemingly insensitive to physical pain	41	2.52	1.63		
Lacks awareness of the existence of the feelings of others	41	2.80	1.54		
Lack of or poor range of emotions	35	2.49	1.57	4.00	0.046
Absence of imaginative play/activity	34	2.53	1.66		

Impairment in initiating or sustaining a conversation with others despite adequate speech	33	2.72	1.40		
Lack of verbal communication with no attempt to utilize an alternative purposeful system to communicate	31	2.29	1.60	4.32	0.038
Repeats or says the same thing over and over	29	2.34	1.50	5.30	0.021
No or abnormal seeking of comfort in times of distress	29	2.38	1.50	5.89	0.015
No or impaired imitation	28	2.34	1.54		
Lacks affect	27	2.12	1.42	4.18	0.041
Regards own hands for prolonged periods of time	27	2.28	1.49		
Maintains interactions by repetitive speech	25	2.42	1.42		
Walks on toes or balls of feet	21	2.12	1.35		
Engages in conversation that is not really appropriate for the context	19	2.19	1.43		
Pedantic (lecturing) style of conversation	16	2.28	1.29		
<b>ADHD</b>					
Fidgets with objects	57	3.33	1.52	4.68	0.030
Does not sit still	56	3.25	1.48		
Frequently interrupts others	55	3.36	1.53		
Gives up on a task easily, or does not complete tasks	54	3.29	1.48		
Always moving—is overly active	51	2.99	1.54		
Does not seem to listen or attend when communicated with directly	46	2.80	1.55		
Appears to daydream	43	3.03	1.33		
Does not wait their turn in activities	42	2.82	1.42	5.18	0.023

Loses things	33	2.84	1.37
Forgets things	19	2.33	1.26
OCD			
Needs to have things in a certain order, or symmetrically, or arranged perfectly	36	2.69	1.54
Seeks reassurance about having done or not having done a certain thing	26	2.30	1.49
Needs to touch or tap certain objects or parts of the body, or needs to blink or not blink	26	2.34	1.56 5.83 0.016
Does the same thing over and over again, or needs to do them a certain number of times or in multiples of certain numbers	22	2.23	1.41
Counts to themselves, or counts objects or words or blinks or whatever	16	2.04	1.24
Checks things over and over again	14	1.79	1.22
Sorts through things over and over again before being able to throw them out, or is not able to throw things out because of "needing it some day"	14	2.19	1.27
Washes over and over again, or in certain ways	13	1.92	1.23
Cleans things over and over again	13	1.81	1.27
Says certain phrases or prayers to make everything okay	10	1.86	1.15
Traces letters over and over again when writing to make them perfect, or reads the same thing over and over again in case something was missed	7	1.92	1.12
Makes sure not to step on cracks or walk under ladders or other superstitious types of things	7	1.67	1.09
Checks work over and over again to make sure there are no mistakes	5	1.82	1.03

## DEAFBLIND

Extreme preference for certain toys, people, food, etc.	69	3.78	1.46	9.91	0.002
Stares at light sources	54	3.08	1.70	6.07	0.014
Vocal self stimulation	54	3.12	1.56		
Tactile defensiveness	53	3.12	1.57		
Dependent on prompts/learned helplessness	51	3.03	1.44		
Plays with light	46	2.88	1.68		
Inappropriate vocalizing	46	2.91	1.60	9.71	0.002
Eye pressing	38	2.53	1.62		
Aggressive behaviors	38	2.65	1.42		
Mouths objects	38	2.59	1.62		
Taps on body	37	2.67	1.66	14.08	0.000
Extreme clinging behaviors	35	2.44	1.42		
Self injurious behaviors	34	2.49	1.58	11.49	0.001
Rocks self	33	2.33	1.58		
Lines things up	33	2.54	1.52		
Darts or runs away	29	2.34	1.53		
Taps on objects	27	2.31	1.42	4.72	0.030
Spins in circles	27	2.38	1.49		
Overly submissive	19	2.06	1.20		
Smears feces	16	1.65	1.32	5.59	0.018
Excessive or public masturbation	9	1.62	1.07		

## TIC

Sudden, rapid, recurrent movements such as eye blinking, head jerking, shoulder shrugging and facial grimacing	41	2.65	1.66	4.04	0.044
Sudden, rapid, recurrent vocalizations such as throat clearing, yelping and other noises, sniffing and tongue clicking	29	2.33	1.52	7.76	0.005

\* Chi square analyses comparing those who were and were not deaf-blind

Table 6

*Intercorrelations Between Behavioral Categories*

Category	1	2	3	4	5
1. Autistic		.60	.55	.47	.66
2. ADHD			.36	.42	.57
3. OCD				.31	.49
4. Tic					.59
5. Deafblind					

Note: all correlations significant at  $p < .01$

Table 7

*Correlations with Behavioral Categories*

Variable	Total	Autistic	ADHD	OCD	Tic	DB
Age	.24**	.26***	.21	.23**	.19	.13
Hospital	.12	.12	.11	.10	.07	.09
Surgery	.03	-.01	.06	.03	.01	.02
Stay	.16	.17	.11	.05	.16	.11
Medical	.17	.22**	.10	.06	.21*	.10
Walking	.20*	.17	-.11	.09	.20	.26***

\*p<.05 \*\*p<.025 \*\*\*p<.001 (two-tailed tests)

Table 8

*Mean Difference in Behavior for Deaf-Blind*

Behavior	N	Mean	sd	<i>t</i>	df	sig
Autistic	66	71.18	19.19	2.48	94	.02
	30	60.57	19.97			
ADHD	66	31.01	7.95	1.69	94	.10
	30	27.93	9.04			
OCD	66	27.21	8.34	1.14	94	.26
	30	24.97	10.25			
Tic	66	5.53	3.02	2.66	94	.01
	30	3.90	2.16			
Deafblind	66	58.12	15.58	3.21	94	.002
	30	47.17	15.30			
Total	66	193.77	44.16	2.93	94	.004
	30	164.53	47.96			

Note: subgroups of deaf-blind (N=66) or not deaf-blind (N=30) are compared with regard to total scores on items associated with specific diagnoses