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Behavioral Approaches to Training Developmentally Disabled Children for an Overnight EEG Procedure

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Abstract

Smith-Lemli-Opitz syndrome (SLOS) is a genetic syndrome associated with multiple congenital malformations, mental retardation, and autism spectrum behaviors. This clinical protocol was part of a larger study investigating the effects of a cholesterol-lowering medication for SLOS patients. Behavioral therapists were consulted to facilitate participants' cooperation with an overnight electroencephalogram (EEG). Seventeen children participated in one 1-hour training session of a mock EEG. Behavioral methods included task analysis, differential reinforcement, and escape extinction. Descriptive data reveal low cognitive and adaptive functioning. Fifty three percent of children tolerated all steps of the training procedure and 88% of participants tolerated all of the actual EEG procedure. Behavioral methods of training children may be an effective preparation for EEG procedures for children with SLOS. This study indicates that sedation, anesthesia, or restraints are not necessary to accomplish EEG testing of children with SLOS. Results may generalize to children with a range of disabilities.

Keywords

Behavioral training; EEG; Developmental disability

Smith-Lemli-Opitz syndrome (SLOS) is a developmental disability syndrome due to an inborn error of cholesterol biosynthesis (Tint et al. 1994). SLOS is associated with multiple congenital

malformations (e.g., growth retardation, heart and brain malformations). Cholesterol-altering therapies have been noted to aid in growth status and, anecdotally, have been hypothesized to improve behavior (e.g., reduce irritability). The behavioral phenotypic expression of SLOS is broad, including cognitive deficits ranging from borderline intellectual functioning to profound mental retardation, sleep cycle disturbance, sensory reactivity, irritability, directed aggression, and self-injurious behavior (Tierney et al. 2000). As many as 46% of SLOS patients meet DSM-IV and Autism Diagnostic Interview diagnostic criteria for autism (Lord et al. 1994; Tierney et al. 2001).

The present clinical protocol was part of a larger study investigating the effects of simvastatin, a cholesterol-altering medication, for SLOS patients. All participants of the larger study were required to participate in an overnight scalp electroencephalogram (EEG), to examine the effects of this medication. Behavioral therapists were consulted to provide clinical intervention to facilitate all participants' cooperation with the EEG procedure.

Cooperation with EEG procedures is challenging for individuals with developmental disabilities, particularly for children with autism and mental retardation (Dorenbaum et al. 1987; Mehta et al. 2004). In some settings this difficulty is addressed by employing restraint, sedation, or anesthesia; these methods can alter medical procedure results and cause unnecessary stress for the child. In contrast, behavior therapy techniques have been used to increase children's compliance with a number of stressful medical procedures such as neuroimaging, positive airway pressure therapy, and radiation therapy (Koontz et al. 2003; Slifer 1996; Slifer et al. 2002). No existing studies target SLOS patients specifically for behavioral intervention to increase compliance with medical procedures. However, children with developmental disabilities (e.g., Down's syndrome, mild to severe mental retardation) and premorbid behavioral difficulties (e.g. hyperactivity) share similar characteristics to the behavioral phenotype of SLOS and are among those patients successfully treated with behavioral therapy approaches (Koontz et al.; Slifer; Slifer et al.). The objectives of the current presentation were to evaluate the efficacy of a brief, behavioral intervention to increase the compliance of children with a developmental disability with an overnight EEG procedure.

Method

Our study was performed in compliance with the Code of Ethics of the World Medical Association (Declaration of Helsinki) and standards established by the Institutional Review Board (IRB) of the Johns Hopkins Medical Institutions and Autism Speaks. All of the behavioral training procedures used in this study were specifically described in the IRB application for the simvastatin study and approved as a part of that larger study.

Behavioral training was provided to 17 children. The treatment protocol was developed and implemented to increase compliance with the overnight EEG procedure. Behavioral methods included task analysis, differential reinforcement of appropriate behaviors, and escape extinction.

Descriptive Measures

Stanford-Binet Intelligence Scales—Fifth Edition (SB-FE; Roid 2003)

The SB-FE is a standardized test designed to assess cognitive abilities in children and adults aged 2–23 years. The SB-FE measures cognitive abilities across four areas, and includes a composite intelligence score. The SB-FE has demonstrated good reliability and validity (Roid). The composite score was used to describe cognitive ability in this sample.

Vineland Adaptive Behavior Scales—Second Edition (Vineland-II; Sparrow et al. 1984)

The parent/caregiver rating form of the Vineland-II is a semi-structured interview designed to assess personal and social skills of daily living. The Vineland-II yields four factor scores and an adaptive behavior composite (ABC) score and has demonstrated good reliability and validity (Sparrow et al.). The ABC score was used to describe adaptive behavior functioning in this sample.

The Sensory Profile (SP; Dunn 1999)

The SP is a caregiver report questionnaire designed to evaluate response to sensory events in children aged 3 to 10 years. This measure yields scores for the modulation of sensory input, and behavioral and emotional responses to sensory stimuli. Lower scores indicate poorer performance. The SP has demonstrated good reliability and validity (Dunn). The multi-sensory processing and social-emotional response scores were used to describe responses to sensory events.

Procedure

A task analysis was developed for the EEG procedure to facilitate teaching of the steps of the EEG procedure to children (see Table 1). Using this method, the EEG was broken down into small, component steps to facilitate training by allowing children to master each individual step of the procedure, while gradually and cumulatively working up to completion of the entire EEG. The task analysis was developed by the senior author and colleagues based on clinical experience teaching children with disabilities to cooperate with medically necessary EEG recordings (e.g., for diagnosis of seizure disorders or for overnight studies for diagnosis of sleep disorders such as sleep apnea). The task analysis steps were selected based on discussion with EEG technicians and by direct observation of cooperative children being prepared for (e.g., electrode placement) and undergoing EEG recordings. Children receive positive reinforcement at the completion of each individual step of the task analysis. No formal stimulus (reinforcer) preference assessment was conducted. The reinforcers were selected, based on parent's report of the child's most preferred stimuli, activities, foods, etc. These were presumed reinforcers based on the Premack principle that high probability behavior can be used contingently to increase the occurrence of lower probability behavior. Specifically, preferred stimuli such as stickers or bites of favorite foods were delivered on a continuous schedule of reinforcement (CRF) after the child cooperated with each step of the task analysis. A larger, tangible "prize" was awarded (fixed ratio-FR12) at the end of the chain of behaviors from the task analysis (following a forward chain sequence). Previous research has demonstrated good interrater reliability with a data collection technique similar to the current task analysis (Tucker et al. 2001). Due to the schedule for the research protocol and limited access to participants (e.g., families traveled to participate, arriving in town the same day this training occurred), training sessions occurred on the day immediately prior to the actual EEG, and were limited in length to one 1-hour training session. Secondary to this time limitation, the mock EEG training protocol consisted of application of 12 EEG electrodes to the child's head, while the actual EEG required placement of 21 electrodes. The use of only 12 electrodes was consistent with clinical experience that children trained to cooperate with 12 electrodes typically are successful with the full array of electrodes during the actual EEG.

During the mock EEG, demands were implemented by verbal instructions and modeling (therapist demonstrated the application of EEG electrodes to a stuffed animal). Therapists provided differential reinforcement to shape instruction following and tolerance of the application of the electrodes; preferred activities (e.g., cartoon movie) and snack items were provided as reinforcers and were delivered on a CRF schedule when compliant behaviors were exhibited during the practice session. As part of the differential reinforcement of appropriate

behaviors (DRA) procedure, reinforcers were withheld contingent on behavior that may disrupt the acquisition of EEG data (e.g., blocking the placement of or removing electrodes). This blocking was also conceptualized as a form of operant extinction of escape-motivated behavior. Therapists ignored negative vocalizations (operant extinction), provided verbal prompts to perform appropriate behaviors, and redirected children to the pleasant activities. Escape extinction was employed wherein children were prevented from escaping unpleasant task demands. Specifically, children were prevented from removing electrodes from their heads via “hands-down” verbal commands and, when necessary, gently blocking them from reaching their heads. Children’s accompanying family members were trained in these behavioral methods during the mock EEG session.

During the actual EEG, children were provided with access to the same pleasant activities and food rewards that they showed preference for during the mock EEG. As much as possible in working with the EEG technician, during the actual EEG procedure the family member and a research assistant provided the same reinforcement and extinction contingencies prompts that had been used during the mock EEG. The overnight EEG was scheduled to begin around the child’s bedtime, to facilitate readiness for sleep onset.

This paper represents a data-based outcome study on a sample of youth who were participating in a longitudinal simvastatin study. As such, the outcome data reported here are not the results of a controlled experimental study of behavioral procedures. Thus, there was no experimental design for the behavioral training procedures reported here.

Results

Participants consisted of 10 males and 7 females ranging in age from 4 to 17 years old ($M=9.3$, $SD=2.6$). Descriptive data on cognitive, adaptive, and sensory processing functioning is summarized in Table 2. Compared to normative group data, the children in this sample scored, on average, in the lowest possible categories on both cognitive and adaptive testing (i.e., “extremely low” and “low” categories for cognitive and adaptive testing, respectively). Multi-sensory processing and emotional-social responses scores were similarly below average as compared to scores of typically developed children.

Fifty three percent of children ($n=9$) tolerated all steps of the training, with the remaining participants ($n=8$) tolerating 75% of training steps. A series of Pearson product-moment and point-biserial correlations revealed no significant relation between descriptive variables (e.g., age, gender, cognitive score) and toleration of training. During the actual EEG, all but two of the participants (88%, $n=15$) tolerated placement of all required 21 EEG electrodes; the remaining 2 participants each tolerated placement of 9 electrodes (resulting in a reduced amount of EEG data). Both participants that tolerated only a reduced placement of EEG leads were noted to require significant amounts of guided compliance in training sessions, secondary to global compliance difficulties (e.g., failure to follow simple commands, such as “sit in chair”). EEG data were collected, on average, for over 9 h for each participant (M in minutes = 577.1, $SD=144.4$), exceeding the desired minimum of 6 h of EEG data.

Discussion

Behavioral methods of training children, via use of a task analysis, differential reinforcement, and escape extinction may be an effective preparation for EEG procedures for children with SLOS. Due to time constraints related to the 1-hour training session on the day prior to the actual procedure, therapists were not able to train children to 100% compliance prior to the actual EEG. In spite of this limitation, the vast majority of children were able to tolerate all steps of the actual EEG procedure, which is noteworthy as almost half the sample tolerated

only 75% of steps during the training session. It is possible that the children who were unsuccessful experienced a deterioration of performance in the latter parts of the EEG set-up because they had only been trained with 12 or fewer electrodes during the mock EEG. This discrepancy may have created a contrast effect when the child was confronted with the full array of 21 electrodes. Based on findings from other studies (Koontz et al. 2003; Slifer 1996; Slifer et al. 2002) regarding the length of procedural training required with developmentally disabled youth, compliance with the EEG procedure may have increased in this sample with the addition of more training sessions. Due to similar behavioral phenotypic characteristics among SLOS patients and individuals with other developmental disabilities (e.g., autism, Down's syndrome), these clinical outcomes may generalize to children with a range of developmental disabilities.

The current clinical protocol is limited by lack of a control condition or group to which to compare results and evaluate intervention efficacy. Another possible limitation of the current protocol is the unknown and potentially confounding effects of the medication being investigated in this study. Due to the double-blind nature of the larger experiment for which the behavioral protocol was designed, it was impossible to identify which participants were prescribed, and possibly deriving benefits from, the medication. Experimental research should be conducted to systematically evaluate the efficacy of this clinical intervention. At minimum, this study indicates that sedation, anesthesia, or restraints are not necessary to accomplish EEG testing of children with SLOS.

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Table 1

Task analysis of EEG procedures for behavioral training

Step	Compliance? Yes = √	Escape/Avoidance Behavior? Yes = √
Enter Room		
Get onto bed/chair		
Touch head		
q-tip on head		
q-tip with NUPREP on head		
Place 1st lead		
Touch head		
q-tip with NUPREP on head		
Place 2nd lead		
Touch head		
q-tip with NUPREP on head		
Place 3rd lead		
Touch head		
q-tip with NUPREP on head		
Place 4th lead		
Touch head		
q-tip with NUPREP on head		
Place 5th lead		
Touch head		
q-tip with NUPREP on head		
Place 6th lead		
Touch head		
q-tip with NUPREP on head		
Place 7th lead		

^a Above task analysis was condensed for purposes of this presentation; task analysis continues in this manner until 12 leads are placed

Table 2

Participant cognitive, adaptive, and sensory processing characteristics

Variable	Mean	SD
Stanford-Binet Composite Intelligence ^a	51.67	8.44
Vineland Adaptive Behavior Composite	46.93	15.39
Sensory Profile Multi-Sensory Processing ^b	23.25	3.82
Sensory Profile Emotional-Social Responses ^b	63.25	9.01

^aMissing data, *n*=8^bMissing data, *n*=5