

A Behavioral Approach to Eliminate Self-Mutilative Behavior in a Lesch-Nyhan Patient

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Lesch-Nyhan disease is an inherited biochemical disorder characterized by the defective activity of the enzyme hypoxanthine guanine phosphoribosyl transferase. A striking feature of this disease is the unusual pattern of self-mutilation that appears in these patients. The present study attempted to assess the effects of applying a behavioral procedure to control self-mutilative behavior in a child diagnosed as having Lesch-Nyhan disease. The behavioral procedure, reinforcement (attention) for responses incompatible with mutilative behavior, was implemented by three individuals and in several different settings. The results demonstrated that the behavioral procedure was effective in eliminating the self-mutilative behavior of this Lesch-Nyhan child. The effects of the procedure successfully generalized across settings and therapists, and were maintained for at least a seven month period.

The Lesch-Nyhan Syndrome (Lesch & Nyhan, 1964) is a heritable malfunctioning of the metabolism of uric acid in which abnormal behavior is a prominent manifestation. This disorder, which occurs exclusively in males, results from a variant gene on the X chromosome. The molecular expression of the mutated gene is the lack of activity of the enzyme hypoxanthine guanine phosphoribosyl transferase (HGPRT) (Seegmiller, Rosenbloom & Kelly, 1967) which usually converts hypoxanthine and guanine to nucleotides. The defective activity of HGPRT results in an overproduction of uric acid, and in turn the presence of excessive uric acid may result in the development of symptoms of gout. All patients with this disease exhibit additional neurological abnormalities which include spastic cerebral palsy and involuntary choreoathetoid movements. Abnormal movements are usually prevalent at an early age and are intensified by tension or excitement. Opisthotosis or extensor spasms of the trunk are characteristic. Also mental retardation is frequently seen in these patients; however, there is some question as to the validity of using IQ tests with these children.

One of the most prominent features of a Lesch-Nyhan patient is self-mutilation. Self-mutilative behaviors may include: biting one's mouth and/or lips, poking one's eyes, picking one's mouth with one's fingers,

scalding oneself in hot water, or catching oneself in the spokes of a wheelchair. As Nyhan (1976) states, this mutilative behavior is characterized by its lightning-like rapidity. When out of restraints, the hand my instantly go to the mouth. It is amazing that patients who are so athetoid, that is, who have such severe difficulty in controlling the simplest of movements, can, so rapidly and so accurately, move their hands to their targets (Nyhan, 1976). Thus, the main control for self-mutilative behavior is physical restraint.

The behavioral manifestations of this syndrome are prevocative, particularly since this disease involves a stereotyped pattern of human behavior that is associated with a specific biochemical aberration. Thus, experimenters have attempted to modify the behavior of such patients by applying chemical therapy. Patients given 5-hydroxy-tryptamine showed a decrease in mutilative behavior; however, the decrease lasted only for a brief span of time (Castells, Chakrabarti, Winsberg, Huric, Perel & Nyhan, 1979). Allopurinol is the chemical applied to lower the concentration of uric acid in the blood, thereby preventing arthritis, renal stones, and fatal nephropathy. Even so, allopurinol has not affected the neurological, cerebral or behavioral manifestations of this disease (Nyhan, 1976). Other investigators suggest that self-mutilative behavior is a learned response, thereby subject to the principles of learning. A few studies have examined the application of behavioral procedures to children with Lesch-Nyhan disease (Anderson, Dancis, Alpert & Hermann, 1975, 1977; Duker, 1975; Nyhan, 1976). Self-mutilative behaviors exhibited by these patients increased when an aversive technique (shock) was applied contingent upon the occurrence of the self-injurious behavior (Anderson et al., 1977). Positive reinforcement (attention, stroking) given when mutilative behavior occurred also increased the frequency of such behavior (Anderson et al., 1977; Nyhan, 1976).

Ignoring biting behavior in a Lesch-Nyhan patient without restraints was reported to be successful in reducing the frequency of that behavior (Duker, 1975). However, this patient was also a head banger and the extinction procedure (ignoring the behavior) failed to decrease that behavior; instead head banging increased. Anderson, et al. (1975) reported that biting behavior in Lesch-Nyhan patients decreased when the experimenter turned away from the patient upon occurrence of the behavior. Nyhan (1976) suggests that the reports so far are only slightly encouraging. However, results obtained in a more recent study by Anderson, et al. (1977) are indeed encouraging. These authors found that positive reinforcement for non-self-mutilative behavior reduced the occurrence of mutilative behavior. Also time-out from reinforcement for self-injurious behavior and time-out plus positive reinforcement of non-self-injurious behavior were most effective in decreasing the frequency of

such behavior.

These studies suggest that self-mutilative behavior exhibited by Lesch-Nyhan patients may be subject to some modification by reinforcement (praise, attention and aversive techniques (shock and time-out). A complementary behavioral technique may be to provide reinforcement for behavior that competes with engaging in mutilative behavior. Indeed, behavioral studies have demonstrated effectiveness of reinforcing incompatible responses to eliminate disruptive behavior in the classroom (Ayllon & Roberts, 1974; O'Leary & Becker, 1967) and to decrease hyperactivity (Ayllon, Layman, Kandel, 1975). Having the patient engage in a competing response limits the chances of the patient exhibiting other undesired behaviors, and, therefore, appears to be a most desirable technique to use with Lesch-Nyhan patients.

The present study attempted to determine the effects of reinforcing incompatible responses (behavior that requires the patient to use hands and thereby competes with mutilative behavior) upon the occurrence of self-mutilation behavior in a Lesch-Nyhan patient.

Background Information

Bobby, the patient, was a 14 year old male, and a resident at an institution for the mentally retarded. He is currently on a unit for mildly retarded, non-ambulatory children. However, this young man had an IQ well within the normal range as indicated by the Peabody Picture Vocabulary test and the Binet intelligence test given in 1972. Also his receptive and expressive language was well developed.

Bobby was diagnosed as having Lesch-Nyhan disease and the collateral neurological manifestations, which include athetotic quadriplegia and cerebral palsy. His seizures were adequately controlled through medication.

Bobby's father is an assistant professor at a small college and his mother, a former employee of the Red Cross, is a housewife. He has two sisters, ages 10 and 9.

Prior to the discovery that Bobby suffered from Lesch-Nyhan disease, he had been in various institutions due to his family's difficulties in dealing with his physical handicaps at home. At the age of three, he was placed in an orthopedic hospital for about two years. At this time he could not walk or hold his back erect so he wore a back brace. Also, leg braces were prescribed and he wore them for two and one-half years. When he was almost six years old, he was placed in a hospital for children with cerebral palsy. After six months he was moved to a school for the handicapped which he attended during the afternoons. In the mornings Bobby attended a typical first grade; however, his parents were

advised that he could not stay in the regular classroom since his drooling was annoying to others and since he had to be taken to the bathroom when necessary. As a result, he attended the first grade for only one month. He was then placed in a pre-school for developmental and learning disabilities. A pediatrician at this institution suggested that Bobby had Lesch-Nyhan disease, following the detection of crystals in his urine. He was taken to Duke Medical Center and enzyme studies (HGPRT assays) found low levels of HGPRT activity (less than .004%) in his erythrocytes, confirming that he had Lesch-Nyhan disease. After the diagnosis, he made periodic visits to Duke for further tests which reconfirmed the original diagnosis.

Collateral behaviors that Bobby exhibited which also reconfirmed the diagnosis included: biting his lip and tongue, injuring his mouth with his hands, biting the inside of his cheek, gouging his tongue frenulum, and throwing himself out of his chair. Also, during an examination he poked the examiner in the eye with his finger. To avoid further mutilative behavior he was placed in arm restraints. Bobby was six years old at this time. The constraints consisted of two arms' lengths of plastic which were strapped together on each arm, preventing the arm from bending and Bobby's fingers from coming in contact with his face.

Additional treatments that Bobby had received for Lesch-Nyhan disease included being given allopurinol to lower the concentration of uric acid and 5 hydroxy-tryptamine to ameliorate self-mutilation. "Unfortunately the drug proved totally ineffective in altering Bobby's behavior" as stated in the physician's report.

Following the diagnosis of Lesch-Nyhan disease, Bobby was moved from the developmental and learning disabilities school to a school for the handicapped. As this move did not help his condition, he was finally put in an institution for the mentally retarded despite his normal IQ and language skills.

Psychiatric reports concerning his progress in the institution stated that if his arm restraints were removed for even a few seconds, he would chew on his hands, and that he exhibited numerous scars on his head, mouth, and chin resulting from self-mutilative behavior. Curiously enough, Bobby believed that he could not control his mutilative behavior and as a result, he, himself, had asked the staff not to remove his restraints. He even dictated a memo to the unit director asking to have the attendants stop asking if he wanted his restraints off. During the periods when his restraints were removed, 10 minute baths and infrequent swim sessions, Bobby either sat on his hands or clutched them to his sides.

This intelligent, articulate young man, was characterized as being severely immobile and almost totally dependent upon others, i.e., could

not use his arms (restrained) or legs (non-ambulatory). The continuous use of restraints presented an additional legal problem, specifically, the use of highly restrictive methods to control behavior. To explore alternative treatments, the unit psychologist requested the assistance of a behaviorally trained therapist to determine if the child's mutilative behavior could be inhibited without the use of restraints. Contrary to all expectations, these plans met with strong resistance from staff members. The major basis for the staff's unwillingness to free Bobby from his continuous restraints was that in their experience any opportunity to move freely had proved to be physically injurious to Bobby. Only after several meetings, discussing and ventilating the staff's feelings, did they consent to allow the restraints to be removed and then only under the supervision and responsibility of the unit psychologist. Bobby's parents were informed of the proposed treatment plan and they enthusiastically gave their consent.

Method of Evaluation

Typically, measurement of behavior prior and subsequent to therapy is the basis used to evaluate treatment effects. In this case, a baseline of Bobby's self-mutilative behavior would have required that he be freed from his restraints. Because of the high risk involved in obtaining this baseline (e.g., the child might poke his eye and blind himself), the evaluation chosen here rested on the known inhibiting characteristics of full restraints versus the yet to be determined inhibiting effects of behavior therapy. Even if each of these two methods proved to be equivalent in effectiveness, the choice of which one to use might be based on the freedom of expression and opportunity for growth afforded the child by one or the other method.

Response Definition and Scoring

Self-mutilative behavior was defined as Bobby picking the corner of his mouth with either index finger and ripping the skin causing his lip to bleed. The length of time he was unrestrained plus any incidents of mutilative behavior were recorded. Also the number of finger to mouth responses (even if he did not rip his lip), of crying, and of requesting to have his hands held when out of restraints was recorded.

Behavior Therapy and Results

The behavioral procedure, reinforcement for incompatible behavior was implemented following the removal of Bobby's restraints. This

therapeutic tactic is based on the notion that a maladaptive behavior pattern can be "displaced" by substituting it with a healthy one. A competing pattern can be increased through the use of reinforcement in a structured situation. Specifically, competing responses in which Bobby engaged included: drawing, throwing plastic darts, playing games, doing puzzles, eating candy, playing catch, reading while holding the book, adding and subtracting numbers on dice, typing, wheeling his wheelchair, and learning sign language. Attention (reinforcement) for engaging in competing responses was applied when Bobby was unrestrained. The personnel included the psychologist, the behavior therapist, and initially the physical therapist.

During the first session the restraints were removed for 26 minutes and, surprisingly, no self-mutilative behavior occurred. At the start of the second session the physical therapist stated that she was very nervous and that she would only remove one of the two arm restraints. The second session lasted for 22 minutes and, again, no mutilative behavior was observed. During both of these sessions, Bobby put his hand to his mouth twice, whereupon, the physical therapist grabbed his hand and put it down from his face. Bobby also requested to have his hands held, twice during the first session and once during the second. The physical therapist was reluctant to continue participating in further sessions, so for sessions 3 (21 minutes) and 4 (23 minutes), the psychologist and the behavior therapist interacted with Bobby and still no mutilative behavior occurred.

During the fifth session, the behavior therapist alone interacted with Bobby for 50 minutes with his restraints off. Not once during this session did Bobby attempt to put his hands to his face, but rather he spent his time manipulating pieces to a puzzle, playing catch, and generally engaging in hand-on-objects activities.

These dramatic results led some of the staff to believe that the child was no longer self-mutilative and hence, a social worker decided to teach Bobby how to feed himself by removing one restraint during lunch. She did so without using the behavioral procedure employed during his therapy sessions. The hour prior to the scheduled 6th session of behavior therapy, the social worker took Bobby to lunch while the behavior therapist inconspicuously observed the interaction. Within 5 to 10 minutes, following the removal of one restraint, Bobby had torn his lip with his index finger, causing his lip to bleed. He cried profusely and immediately the restraint was put back on and the social worker resumed feeding Bobby. An hour later, following the lunch incident, Bobby was conducted in restraints to his scheduled meeting with the behavior therapist. During this session (6th) Bobby was allowed total freedom from his restraints for a continuous period of 3½ hours. No mutilative

behavior or hands-to-mouth behavior was exhibited during this lengthy unrestrained period. Further, since no self-mutilative incidents had been observed at any time during the 5 sessions of behavior therapy, an opportunity presented itself to explore fading the structure associated with competing responses. Specifically, instead of prompting competing responses that required, for instance, that his hands be constantly busy, Bobby was allowed a "free" and unstructured period of time. For example, he spent much of this session (6th) outside on the hospital grounds conversing with the therapist and other passersby.

Because of the marked progress observed in Bobby during session 6, the following day a second attempt was made by the staff to teach Bobby to feed himself while the behavior therapist observed the interaction. Both of Bobby's restraints were removed by an attendant at dinner. Twice during a 15 minute period he engaged in mutilative behavior (tearing his lip with his finger, bleeding, and crying). Again, the child's restraints were put back on and the attendant fed Bobby as usual. The highly inconsistent behavior shown by Bobby when working with staff contrasted to his progress observed in behavior therapy. An examination of the staff's reaction to Bobby's self-mutilative behavior revealed that when the child hurt himself, the staff reacted most sympathetically to his crying and bleeding, thereby unwittingly reinforcing his mutilative behavior, e.g., "Now Bobby don't cry, it's okay."

Since the mutilative behavior was seemingly affected by attention from others, the next step consisted of teaching an additional staff member to react differentially to Bobby's mutilative as well as to his "healthy" behavior when he was unrestrained. Specifically, the special education teacher was directed to: (a) reinforce with social attention whenever Bobby engaged in competing, adaptive behaviors, and (b) minimize giving Bobby undue attention for "sham" attempts at self-mutilation, while at such times prompting him with stimuli (e.g., toys, manipulables) to get him to engage in competing responses. The special education teacher agreed to remove Bobby's restraints during math and reading sessions, to implement the behavioral procedure, and to record its effects. She had previous training in behavior modification and was quite competent in applying the procedures, which made the fading out of the behavior therapist during these sessions easy. Within three sessions, the behavior therapist was faded out. No mutilative behavior occurred during these sessions nor during the next two math and reading sessions when the procedure was implemented by the teacher alone in the absence of the behavior therapist.

One of the major interests expressed by the staff involved teaching Bobby to feed himself. Since the staff had attempted and failed to achieve this objective, it seemed appropriate to first teach such complex

behavior through the psychologist and the special education teacher. Accordingly, the child had his restraints removed during 5 breakfast sessions with the special education teacher. Bobby learned to feed himself in each of these sessions and did so without apparent difficulty. Only one incident of self-mutilative behavior occurred at breakfast during the fourth day. However, Bobby had been upset and crying when the psychologist took him to breakfast. He cried continuously during this time. He stated to the psychologist how earlier that morning a staff member had told him he could never be out of restraints.

Discussion

The results reported in this case study suggest that the Lesch-Nyhan disease may be conceptualized as a behavioral problem susceptible to a behavioral approach. The results of this case study are consistent with previous findings (Anderson, et al., 1975; 1977; Duker, 1975; Nyhan, 1976).

An interesting occurrence during this study was that by the 6th session (3½ hours), Bobby did not engage in mutilative behavior even though his hands were not kept busy. One could surmise that he had learned that if he engaged in mutilative behavior his restraints would be placed on, and that the behavior therapist would only interact with him when his restraints were off. Indeed, Bobby would even ask the therapist not to leave and would say he did not want his restraints placed back on, which would happen after the therapy session was over.

Significant observations were made concerning the relationship between self-mutilative behavior and periods of stress and tension. For example, during math and reading periods, Bobby exhibited worry and concern when he failed to answer correctly but rather than tearing his lip, as was a usual reaction to stress, he learned to touch his mouth with his fingertip without hurting himself. When he answered correctly, his fingertip never approached his mouth. Similarly, meal times had often been stressful since feeding movements were difficult for him due to his cerebral palsy. Even then, he coped with the stress very effectively, using utensils slowly and carefully rather than thrusting the utensils against his person or against others.

In attempting to determine why this procedure, reinforcing competing responses, suppressed mutilative behavior in Bobby, one should analyze the procedure itself. Mutilative behavior exhibited by Lesch-Nyhan patients appears to increase when attention is given for such behavior (Anderson, et al., 1977). Thus the behavior of these patients is affected by social reinforcement. Since reinforcement was given for a response which competed with mutilative behavior, Bobby would have had to stop

the self-competing behavior for which he received reinforcement in order to exhibit mutilative behavior.

To assure generalization of Bobby's "self-control" in the absence of restraints, the therapeutic program required additional personnel to the therapist and additional settings. Accordingly, three different individuals employed the behavioral procedure across different settings: specifically, a male psychologist, a female special education teacher, and a female behavior therapist. Each of them removed the physical restraints from Bobby at meal times as well as during the daily academic and free time periods. At such times, Bobby displayed normal behavior appropriate to the setting. He fed himself at meals in the presence of regular staff and he played with other residents and staff during leisure time. He also followed instructions and completed assignments in the special education room without causing any disturbance.

Seven months after the behavioral treatment, the behavior therapist and another observer visited Bobby at the institution. Bobby was wearing full restraints and the staff reported that he wore them all the time. The therapist took Bobby to a small room and after freeing him for one hour noticed no self-mutilative efforts on his part, despite the fact that a stranger was present at the time.

This study demonstrates that such gains as were made with Bobby were directly related to the treatment used. Further, that in the absence of the treatment, his self-destructive behavior continued unabated. Admittedly, generalization of these therapeutic gains were limited to the settings and individuals where the behavioral treatment was used. It is to be noted that the staff discontinued the behavioral treatment which prevented generalization from occurring. When the visitor met with Bobby, after such a long time, he displayed effective "self-control" over his destructive behavior. This strongly suggests that generalization would have been achieved had the hospital staff continued to adhere to the behavioral treatment, specifically reinforcing competing-adaptive behaviors and minimizing social reinforcement for self-destructive behavior. Theoretically, these therapeutic gains would also have come to be maintained in time by natural contingencies in the environment as contrasted to the programmed contingencies used in the behavioral treatment.

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