A Brief Guide to the Behavioral Care of Individuals Diagnosed with Lesch–Nyhan Disease (LND)

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All of the residents diagnosed with LND who live at Matheny and have taken the time to teach us about what they need and what works.

A Brief Guide to the Behavioral Care of Individuals Diagnosed with Lesch–Nyhan Disease (LND)

A Simple Purpose

The purpose of this guide is to briefly explain the behavioral manifestations of Lesch–Nyhan Disease (LND) and describe how the Matheny Medical and Educational Center has learned to care for this population psychologically and behaviorally over time. Describing to someone what to do is not often sufficient to

elicit a change in behavior, or get a "buy-in" response.

This guide will not only go over what to do, but why to do it and why we believe it works. If you are going to use this please take the time to read it in its entirety, the why is equal in importance to the what.

A Brief Introduction to LND

Lesch–Nyhan Disease (LND), also referred to at times as Lesch–Nyhan Syndrome (LNS), is a highly complex and multifaceted diagnosis. It is a genetic (X-linked recessive) disorder of purine metabolism occurring in roughly one in 380,000 live births (Crawhall, Henderson & Kelley, 1972). It involves the near absence of the enzyme hypoxanthine guanine phosphoribosyltransferase (HPRT).

Persons with classic LND typically have renal dysfunction, movement disorder, dysarthric speech, ocular motor dysfunction, specifically in the controlling of saccadic eye movement, and varying degrees of cognitive impairment ranging from severe deficit to near normal functioning (Matthews, Solan, Barabas & Robey, 1999). The hallmark behavioral symptom of LND is involuntary self- injurious behavior. These behaviors might include: biting of fingers or lips, possibly to amputation, head-banging, eye-poking, and scratching

of self or others (Anderson and Ernst, 1994; Robey, Reck, Giacomini, Barabas & Eddey, 2003).

The involuntary self-injury manifests differently and uniquely in each individual with the diagnosis. Thus, while there are common behaviors, it would be incorrect to assume that an individual diagnosed with LND will inevitably develop any one particular behavior. Very often, the behaviors discussed and focused on are the physical self-injurious behaviors. While these are of paramount concern for physical safety, the emotional and

social self-injurious behaviors can be equally damaging and at times more complicated to safeguard.

Individuals with LND typically use and appreciate a range of protective devices such as wrist cuffs, arm splints, bed bumpers and night time tie downs to safeguard them from the involuntary physically self-injurious behaviors. Dental extraction is sometimes used to protect individuals with LND from self-injury due to biting. The use of safe-guarding measures and equipment are highly dependent on families and diagnosed

individual's comfort, physician and therapist comfort, state regulations and laws, and facility regulations and policies.

Truly Without Control

The self-injurious and self-sabotage behaviors of individuals with LND can be complicated, intricate, and appear manipulative. They have all the signs of planned and controlled behavior; however, they are in fact not under the individual's control. At Matheny we refer to such behaviors as "LND behaviors" for conversational convenience. The truth is they are simply behaviors based on thoughts and emotions just like any other behavior.

The difference is, we get the opportunity to select out the behavior we wish to act on from a myriad of thoughts and emotions, and they do not. Imagine if you acted on every one of your worst thoughts in your most emotional moments; what would your behaviors look like under those circumstances?

For example, picture yourself walking into a DMV, the person behind the desk tells you they lost all of your information, and you need to start the entire process of getting a renewed driver's license from scratch.

It might briefly pass through your brain that you would like to curse at the person, perhaps spit at them, and perhaps even hit them. You take for granted, as we all do, that you get to have thoughts like that and not act on them. You are also aware of the fact that the employee in front of you did not lose your information and this situation is not their fault, but the emotion will be displaced onto them simply because they are there with you. This is a common emotional defense mechanism often referred to as "shooting the messenger." In other words, simply because the person in

front of you is not the cause of your emotional state, it does not mean your behaviors will not end up directed at them. We all have this emotional experience, but we get the opportunity to decide not to act on that emotionality.

Now imagine you did hit the person, and in the next DMV you hit another, and so on, until you get arrested for it. The next time you walk into a DMV what will you be thinking? More than likely you will be begging yourself not to hit anyone, that worrying about your own behavior

causes anxiety and fear, it is that emotional experience that puts you at more risk for the very behavior that caused the emotion. This is the LND cycle.

The previous example gives you a very general depiction of how one might apply a lack of control over our emotional behaviors to a situation most can identify with. However, it is in and of itself misleading because it also alludes to behaviors being intentionally directed, which is not the case for individuals diagnosed with LND. It is a highly imperfect

example to discuss what it might feel like to be afraid of one's own behaviors, and have that fear impact your future behaviors. This illustrates the complexity of describing behaviors related to LND to those of us who do not have LND.

There are rare example that give us insight into the experience of an urge to self-harm which is not based in or fueled by a desire to self-harm. One such instance is what the French have termed l'appel du vide, which translates to English as "the call of the void." This is when a person

confronted with a high ledge, balcony, window, or cliff experiences a sudden urge to jump. This urge can cause some of these individuals to develop a fear of being in high places because they worry that they may compulsively jump.

In a study by Jennifer Hames et al (2012) researchers found that more than 50% of the 431 individuals interviewed experienced this urge to jump. This phenomenon was common regardless of the individual's history of suicidal ideation. This is an example of a

somewhat common urge to put one's self in a situation that would cause serious injury or death without any desire to self-harm or die. It is likely that a substantial percentage of people reading this have experienced this phenomenon. Imagine if that impulse to jump led to you actually jumping without control over the behavior, imagine the fear you would experience any time you confronted a high space.

When individuals with LND find themselves stuck in that thought, emotion and behavior cycle they

require intervention to break it. At times, this may be direct intervention for the behavior, such as safe-guarding or giving a break from an activity. Other times it requires an emotional intervention, such as helping that person find a reason to laugh, directing them to something that helps them feel calm, or making sure they feel truly understood. Finally, there are times in which thought intervention is required, to stop the negative self-talk which would result if any person lost control over their behaviors and did something they regret. This can be

the most difficult part of the cycle to stop. Before discussing the impactful strategies it is important to first understand what is happening in the brain to create this cycle.

Brain Basics

There is a large amount of evidence that LND is, for behavioral purposes, a disease of the Basal Ganglia (Visser, J., Bär, P., & Jinnah, H., 2000). The Basal Ganglia is a part of your brain that is heavily connected to, if not part of your Limbic System. The Limbic System is an interconnected grouping of brain structures that control all of your emotions, positive and negative. In simple terms, this system is responsible for your ability to inhibit negative thoughts that come with

high levels of emotions (Hikosaka, O., & Isoda, M., 2010 & Florio, T., Scarnati, E., Rosa, I., Di Censo, D., Ranieri, B., Cimini, A., Alecci, M., 2018). The specific part of the Basal Ganglia responsible for inhibiting is called the Globus Pallidus (Gittis, A., Berke, J., Bevan, M., Chan, C., Mallet, N., Morrow, M., & Schmidt, R., 2014). This inhibition system, the Globus Pallidus, does not function well in individuals with LND. Therefore, when they become emotional, be that through fear, anxiety, concern, frustration, anger,

or even excitement, they lose the ability to inhibit behaviors.

When doctors have used deep brain stimulation (DBS) to force the Globus Pallidus to function, or increase in activity, the behaviors decrease significantly and in some cases all but disappear (Deon, L., Kalichman, M., Booth, C., Slavin, K., & Gaebler-Spira, D., 2012). According to researchers the individual they treated using DBS demonstrated "*remarkably good results in both dystonia and self-injurious behavior such as those*

achieved by other authors using the same modality. The stimulation benefit remained during the five-year follow-up and still continues today" (Piedimonte, F., et al. 2015).

It is important to state that currently DBS is complex and expensive, and for many struggling with this diagnosis is not a solution they have access to. While this does provide hope for the future, especially as we see more widespread use of DBS for those diagnosed with Parkinson's Disease, this is not why it is discussed in this guide. Instead, it is mentioned

to demonstrate that there is a neurological dysfunction at the heart of the self-injurious behaviors. It is important for those who care for an individual diagnosed with LND and the individual themselves to accept this. Not doing so leads us all to ascribe intent to the behaviors which does not exist, and no benefit will come of that.

Researchers utilized microinjections of bicuculline in the sensorimotor territory and the limbic territory of the external Globus Pallidus (GPe) of primates to study the impact of

damage to and diminished activity in the GPe (Grabli D, et al. 2004). Bicuculline is an antagonist, or a blocker, for the primary neurotransmitter active in the GPe. In other words, it shuts down the activity in that area of the brain for a period of time. This allows researchers to see what happens to behaviors or skills when that part of the brain has diminished activity.

The researchers found that bicuculline microinjections induced behavior disturbances in the limbic part of

the GPe. The expression of behaviors included abnormal repetition of licking or biting of the tail or fingers, and licking the cage bars, none of which were observed outside the effect of the bicuculline microinjections.

In short, the researchers saw some tic-like, and at times self-injurious behaviors, when functioning in specifically the part of the GPe connected to the limbic system was diminished. The researchers state "the behavioral effects shared similar features with symptoms observed in

Tourette's syndrome, attention deficit/hyperactivity and compulsive disorders. Thus, our study provides experimental evidence for the involvement of the associative and limbic parts of the basal ganglia in these pathologies. These results may provide the basis for a primate model of these disorders" (Grabli D, et al. 2004).

It is of paramount importance for those who work with individuals diagnosed with LND to truly accept that the person cannot control their behavioral manifestations,

particularly when they are emotional. The purpose of this section is to illustrate the neural mechanism that explains why someone would not have the control the rest of us have come to take for granted. It is rarely sufficient when asking someone to accept an extreme premise to use the arguments “because I said so,” “just trust me,” “or because science says so.” While this rationale may be somewhat complex, it is important to comprehend the reason the individual with LND does not have control over their behaviors.

Safe-Guarding

Due to the dangerous and at times extreme nature of the physically self-injurious behavior which can manifest from LND safe-guarding devices are often a necessary part of keeping the individual safe. Families and diagnosed individual's comfort, physician and therapist comfort, state regulations and laws, and facility regulations and policies all can impact decision making when it comes to utilizing safe-guarding equipment. Taking all of those variables into account, maintenance

of physical safety for an individual diagnosed with LND is paramount. These are not behaviors that will disappear on their own, that can be behaviorally trained out without equipment, or that can be medicated to extinction.

Safe-guarding equipment should also be designed and consulted on by a physician, physical therapist, and occupational therapist to be sure they maintain safety without causing harm of their own. Safe-guarding equipment can include but is far from limited to: specially designed cuffs

for arms or legs, chest harness, arm splints, as well as modified versions of each of these for lying in bed in different positions. Any equipment such as this must be monitored regularly as it can come lose, or be used by the individual's LND behavior to rub against or cut off circulation.

In some cases involving sever oral damage caused by biting behaviors teeth removal has been utilized. As with safe-guarding equipment families and diagnosed individual's comfort, physician and therapist

comfort, state regulations and laws, and facility regulations and policies all can impact decision making in this case.

Physical safe-guarding responses should be tailored to the specific behavioral manifestation of the individual diagnosed with LND. A safe-guarding device or intervention that is effective and vital to the care of one individual could be dangerous for the behavioral manifestations of another.

When safe-guarding equipment and intervention are utilized appropriately it typically has a calming effect as the individual is no longer living in constant fear of their own behaviors to the degree they do without the intervention. It is important to mention that it is common for social and emotional self-injurious behavior to increase, especially early on, when appropriate physical safe-guarding interventions are put in place. The self-injurious behavior is still present, but manifests differently when they are kept physically safe. This is when the proper treatment of the emotional

and social self-injurious behaviors becomes vital.

Feeling Understood

Individuals with LND are heavily reliant on those around them to keep them safe and to be understanding of their behaviors. If the individuals around them do not do what they need to, the person with LND knows they are now at risk from themselves. A caregiver cannot keep safe an individual they are afraid of, do not understand, or do not like.

This is often what individuals with LND look for in others: understanding of the behaviors that

come with the diagnosis, a lack of fear, and to be liked and seen through their behaviors. If they do not feel those things they cannot develop trust, in the absence of trust fear and anxiety will grow in the presence of that caregiver, that increase in fear and anxiety will cause an increase in involuntary behaviors.

It is important to understand the difference that lies between being understood and feeling understood, between understanding someone and showing understanding. The following strategy, termed selective

ignoring with redirection, is the mechanism we use to demonstrate our understanding of LND to the person who is diagnosed. When someone with LND experiences behaviors, it is easy for anyone to become focused on themselves and the impact the behaviors have on them. If the behaviors are increased, it is safe to assume that something is wrong, showing concern will help demonstrate to the person with LND that you see their struggle. Feeling seen is powerful, selective ignoring with redirection is a vital tool in

showing someone with LND they are seen and understood.

Selective Ignoring With Redirection

At Matheny we use a technique called selective ignoring with redirection to help individuals with LND feel understood and safe. Selective ignoring with redirection means the caregiver completely ignore that the negative behavior occurred at all; if the person apologizes you briefly accept it once, and then redirect them back to a completely unrelated conversation.

Therefore, if someone with LND spits in your face, you don't flinch, you

don't blink, you don't wipe it away in front of them, you don't comment on it, and you don't make a sound. For all intents and purposes you have no reaction to it whatsoever. If they apologize you say, "it's fine, don't worry about it," then immediately transition to another unrelated conversation about something they like, are interested in, a shared interest, or anything else that is appropriate. This reaction is the same if they say something sexist, sexual, racist, against your religion, family, or any negative behavior. You are a human and may feel understandably

grossed out or offended, but that reaction should be experienced later, by yourself, outside of earshot and line of sight and not when you are with the individual.

There has been research on LND as a movement disorder, a genetic disorder, a kidney disease, and as a neurological disorder; however, there has been little research on LND as a psychological and behavioral disorder. There was a study done by Lowell Anderson, Joseph Dancis, and Murray Alpert out of the New York University (NYU) School of

Medicine in 1978 on LND as a behavioral condition.

This group studied the therapeutic effectiveness of punishment, positive reinforcement of either self-injurious behavior (SIB) or non-SIB, and time-out learning paradigms. They found that "*positive reinforcement of non-self-injury and time-out from social reinforcement were consistently and rapidly effective, indicating a complex interaction of genetic and environmental factors in the production of SIB*" (Anderson, L., Dancis, J., & Alpert, M., 1978).

What they are describing is the first step toward selective ignoring with redirection.

Anderson et. al. further found that "*The response prevention procedure was evaluated four times during the 6-day test. On each occasion there was a consistent and relatively stable rate of between four and seven attempts at self-injury per minute. When shock was administered without interrupting the behavior (response prevention discontinued), the self-injury rate remained at about the*

level of the response prevention "baseline." When shock and response prevention were combined, the rate increased to almost twice that of either procedure alone. Time-out produced a rapid decrease in SIB to less than one per minute" (Anderson et. al., 1978).

What researchers are talking about here is that introducing a negative to stop the behavior, which is punishment, was not only ineffective but it actually made the behavior significantly more likely to occur. Extrapolating on their findings, this

means that if an individual with LND is scolded for behaviors they exhibit, that behavior will more than likely increase in frequency and intensity not decrease, especially in the presence of the person doing the scolding.

There may be behaviors which threaten the safety of the individual or the safety of another that cannot be completely ignored. Selective ignoring with redirection does not mean allowing the person to harm themselves without intervention. There are likely to be instances of

minor physical harm that can be ignored and redirected. There may be other instances of harm or risk of harm that demand physical intervention.

Individuals with LND often require the intelligent and careful use of safe-guarding equipment. If equipment became loose or required adjustment or if the individual were to get free of their safeguarding equipment you would physically intervene to help them reapply it; however, you would have no verbal, emotional or facial reaction to it. The goal would be to

keep the individual physically safe and talk to them about something completely unrelated to the situation while doing so.

Even a statement that may seem benign such as “relax your arm” is not selective ignoring with redirection, because you are drawing attention to the fact that the person’s arm is not relaxed. You will likely note that such statements increase tension in the part of the body which you refer to. Whereas talking about the football game the night before with an individual who enjoys

football while reapplying or adjusting the safeguarding equipment has a much higher rate of success.

Researchers observed three children diagnosed with LND individually, in different contexts of daily life, always with their usual safe-guarding equipment and in the presence of a caregiver. Over the course of 60 observational sessions, for over 90 total hours, the team documented 292 LNBs, interfering with different aspects of life (Bozano, A, Schiaffino, A, Spessa, A, et al., 2020). The team found "*the most*

useful strategies in both harm to communication in progress and harm to an activity in progress are irony and distraction, and physical containment for the latter. The coefficient of scolding is negative in all types of harm, especially in harm to others/objects. Regardless of the strategy used, the management approach of an agitated or upset caregiver seems unlikely to be effective; on the other hand, calmness alone does not necessarily stop the LNB underway."

The researchers found once again that scolding and punishment exacerbate self-injurious and self-sabotage behaviors in individuals with LND and that distraction and redirection are powerful tools. However, what the researchers also describe from their observations, is that the mood of the caregiver is as important, if not more important, than the strategy they use. If a caregiver is noticeably upset or agitated, use of the strategies is no longer effective, regardless of the strategy used. This is why selective ignoring with redirection is more than a prescribed list of actions to take and

actions to avoid. One must truly believe that the individual they work with does not control the behaviors, avoid frustration and blame, and understand why they are selectively ignoring and redirecting.

Selective ignoring with redirection may sound extreme, it may sound difficult to impossible, and it may sound like you are allowing the behavior. This is the most effective technique for this population and works extremely well in decreasing the involuntary behaviors. You may be asking why, which is a fair and

important question that is addressed in the next section.

It Is Not About You

Individuals with LND are always harming themselves. Even when the behavior involves harming you; it is not the final impact of the behavior. If someone with LND spits at you, the fear that brought the involuntary behavior on is most often the thought that you will be grossed out, not want to work with them or keep them safe, and then eventually they will be left alone and in danger. The behavior involves something unpleasant to you, but it is still involuntary self-injury.

When you utilize selective ignoring with redirection you are showing the person with LND, in the most extreme fashion you can, “I understand you do not control your behaviors so they will not drive me away, I will not leave you alone, I will be here to keep you safe.” That gesture decreases their anxiety of their behaviors coming out around you, which decreases the negative emotions in the limbic system, which decreases the load on the mechanisms that inhibit behavior, which means you get less of those involuntary

behaviors. However, the moment someone else walks up who does react to the behaviors, you will see the anxiety and the involuntary behaviors return.

Loss For Words

There are aspects of LND which are difficult to discuss in the English language. We have words for a behavior we have no control over at all, such as involuntary; we have words for a behavior that has become automatic over time and learning, such as habitual; and we have words for behaviors we use to describe those geared toward attaining a desired object or emotion, such as addictive. But, a descriptor for a behavior that we simultaneously are compelled to act on and struggle desperately

against its release without the promise of secondary gain? Such a term does not exist; how often are humans in a position where we need to describe such a behavior?

It may seem odd to discuss terminology when there are so many other challenges that come with this diagnosis, but what we call something is important. If a doctor tells you that you seem sad, depressed, melancholy, or grief stricken, do those terms not change how you feel about your own state? The terms aggressive and violent are very similar in some

contexts; however, if a doctor told you they were going to violently treat your illness you would likely be concerned.

Language of behaviors can assign blame, it can assume intent, it can overlook a struggle, and it can strongly influence how the recipient of that term views themselves. In a journal article entitled "*Wanted: A vocabulary for talking about involuntary behaviors associated with Lesch-Nyhan disease*" Dr. Robey and this writer discuss the issues terminology create when training,

teaching, and working with individuals diagnosed with LND. In this article we suggest the possibility of terming the “LND behaviors”, as we often refer to them at Matheny, “inexorable behaviors” and suggest the utilization of the verb “compelled” to describe those behaviors. “Together, perhaps the verb “compelled” and adjective “inexorable” help to illustrate the internal struggle that individuals with LND have conveyed to us that they experience” (Robey, K.L., Balboni, D. 2021).

It is important to remember when working, or teaching others to work with this population the language you use is important. There is a chasm between being understood and feeling understood. Do your best to use language that conveys your understanding to the individual with LND. Remember, the person you care for is only hoping the people they care about and the people who care about them will see the person underneath the behaviors.

A Difficult Ask

LND is a complex diagnosis and the behavioral description in this guide is a general one. Each individual with LND will have behaviors that manifest differently. Therefore, selective ignoring with redirection will need to be tailored specifically to each individual with LND based on their behavior profile, personality, likes and dislikes. This brief guide is not meant to give the answers on how to work with any individual with LND. It is a roadmap on how to find answers for the individual you work

with. Selective ignoring with redirection is a hard thing to put into practice, and it takes practice and time to do well, as it requires fully accepting that a person does not control their behaviors.

It is important to remember that the interaction is infinitely harder for the person you are working with than it is for you. Put yourself in that person's position. Imagine you cursed at, spit at, hit, and said the worst things you can think of to everyone you love the most in your life; your parents, girlfriend, boyfriend, husband, wife,

kids, and friends. Now imagine that your only hope of maintaining that relationship was that they believed you when you said you could not control your behaviors. From that perspective, it is worse to spit on someone than to be spit on; it is worse to say something terrible than to have it said to you; it is always harder for them. This will help you find the empathy you will need to truly use selective ignoring with redirection.

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