

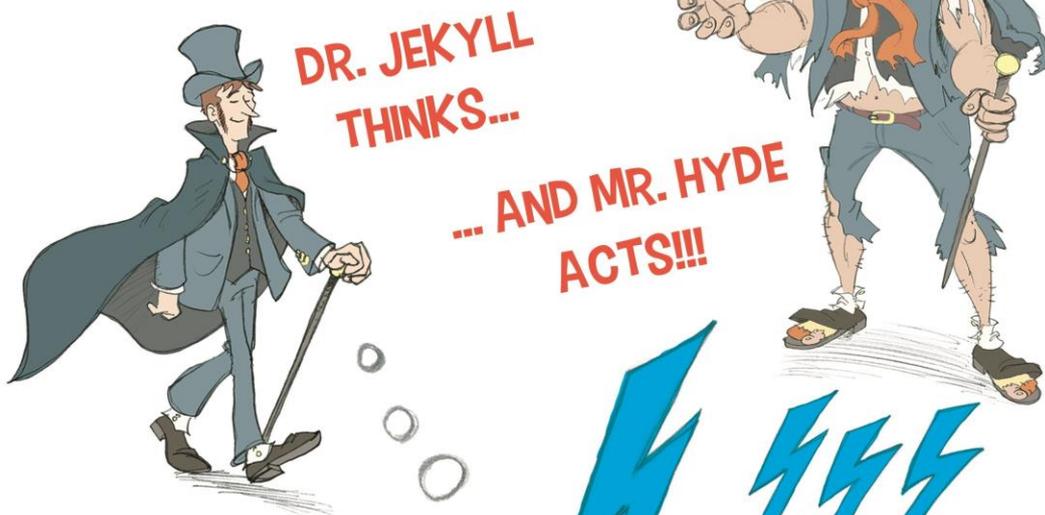
Living with Lesch-Nyhan:

suggestions to manage Lesch-Nyhan behaviours more effectively.

**LESCH-NYHAN SYNDROME:
THE ILLNESS THAT ROWS AGAINST MY WILL!**



I FEEL LIKE DR. JEKILL AND MR. HYDE, SOMETIMES...



I WANT TO CARESS MUM...
LITTLE BROTHER I LOVE YOU...
I'M THIRSTY AND I TAKE A GLASS OF WATER...
I'D LIKE TO BE QUIET...

... BUT MY HAND GIVE HER A SLAP.
... BUT A SWEAR WORD LEAVES MY MOUTH.
... BUT MY HAND THROWS IT ON THE FLOOR.
... BUT MY TEETH BITE MY FINGER, MY LIP OR MY TONGUE.

1. Be aware that LN people do not want to cause damage to themselves, to property or to others

Examples:

- As soon as they meet, D. spits on the other person who responds by saying "Don't worry, I know it's your disorder, I'll wear a raincoat!" The action is not repeated for the whole meeting.

- Some very young children arrive in front of M. who is worried about kicking them and asks that they be kept "at a safe distance"

*It's a waste of time for the caregiver and frustrating for the LN person to want to convince them that an action is wrong, damaging, dangerous, painful: sometimes these issues violate suggestion 9 and actually become damaging. This also applies to the words spoken: **see suggestion 7.1***

2. LN behaviour produces unintentional actions as well as an unintentional incapacity to act.

Examples:

- M. (LN) raises his hand to respond and when asked to speak, says nothing.

- A. (LN) has to push the button to continue playing and remains immobile.

- M. (LN) has some outpatient exams and refuses to urinate. These non-actions often lead to anger.

3. Be aware that the times when the person cannot control themselves are the most vulnerable ones

Examples:

- the orthodontist asks the mother to CONVINCER her son not to spit out the retainer that doesn't fit perfectly...the mother is frustrated because she can't explain that if S. managed to "not spit out the retainer" he would also be able to "not bite his lips" so he wouldn't need the retainer!

4. Be aware that it is possible for people with LN to achieve autonomy through mediation, and by containing and accompanying them

Examples:

- F. (LN) likes painting with his hand in the teacher's hand, A. (LN) likes colouring a drawing,

- M. (LN) likes kneading bread dough or making ceramic objects. M. (LN) likes writing stories or poems even without a keyboard. D.(LN) has fun playing the keyboard for up to 15 minutes even without his protective glove, if he's got someone with him ready for his forewarning.

Being able to express and complete what they want is satisfying and gratifying for these youths, but they can't manage this without constant back-up, without someone who can "hold their hand", without appropriate assistance. They require aid and mediation to achieve their freedom and autonomy.

Helping them become more autonomous does not necessarily mean aiming at self-sufficiency

5. Work a lot on verbal and non-verbal communication

These youths are very attentive and communicative, but the people around them are not always capable of non-verbal communication. Augmentative/alternative communication provides great management possibilities for all youths with serious dysarthria, but with LN youths it is also a possible behavioural management tool

Examples:

- E. (LN) was given the chance to use a symbol on a wristband to indicate when his action was unintentional; when he indicates "I didn't mean to" with his other hand, it seems to give him a great deal of satisfaction and E. expressed his desire to be able to use this tool at school.

- M. (LN) cries because his mother doesn't understand what he wants to say. His mother can't get him to clearly say yes or no because he is conditioned by LNB. She proposes lifting his arm for yes and his leg for no; M. does this, and stops crying and getting distressed. His mother says that 99% of the time, LN doesn't interfere with this system...

6. Keeping a safe distance

Since the actions of LN youths are lightning-fast and powerful, it is fundamental to get used to keeping a safe distance between the LN person's head and the body of the other person, or walls, doors and poles. If LN people do not have their arms and legs tied, then also between them and moving objects near them (e.g. cups of coffee), objects sitting on a table within arm's reach (plates, glasses etc.), nose, stomach, glasses of the other person.

These examples are only give for accidents caused by the distraction of caregivers, so that in this case, the experience of the surroundings is not forgotten.

- eight-year old L. runs towards his LN classmate who he hasn't seen for a while, and in the meantime he takes off his glasses and puts them in his pocket...
- A. (LN)'s kindergarten classmates stop a boy from another class from telling the teacher that A. kicked him because: "you should have been more careful, it's not his fault".
- E. (LN)'s swimming instructor broke their nose because E. suddenly butted their head back.

7. Calmness, patience, restraint, ability to wait

These are essential to understand LN children and youths who struggle to speak and may not be able to say or do what they want to, or even to explain their discomfort if pressed with questions, if the other person yells at them, if they get a feeling of distress, haste, fear.

Examples:

- E. (LN) had to change educator because the one he had always arrived a bit late and was forced to do everything in a rush, which caused psychomotor agitation, confusion, disorganised conduct and emotional distress. The problem disappeared with a punctual educator.
- S. (LN) asked the Emergency room doctor to stop telling him to stay still so he could manage to stay still.
- The silent wait of the support teacher ensures that it is M. (LN), whose finger impulsively pressed the wrong key, who corrects the direction and presses the right letter.
- The teacher asks F. (LN) a question, then after the immediate answer, repeats the question and waits for the second answer, the one given calmly and that indicated the boy's real wishes. "Do you want some more yogurt? " "no" "...yes".

7.1 Often the first answer given in a hurry isn't what was really intended and can be wrong

7.2 Showing high expectations sets the scene for LNB

7.3 Group activities and integration with peers reduce the risk of LNB

7.4 Taking your turn to talk helps manage LNB

8. Don't overcrowd the person with tasks to do at the same time

Examples:

- E. (LN) wants to narrate something while he's in the middle of doing something; the physiotherapist stops him and says "We'll talk later". E. completes the action.
- The rehabilitator tells A. (LN) to press a key, choose a colour, answer LN yes or no, without waiting and A. does none of the above.

9. Don't talk about or mention or use facial expressions to bring up LNB already implemented in the past or potentially dangerous/not correct

Example:

- S. (LN)'s mother was telling someone on the telephone that after she had bandaged S.'s finger, he didn't bite it any more. "Now his finger has healed, we removed the bandage and he doesn't do it anymore." The child heard this and immediately put his finger into his mouth.
- Others: "Now you have to be quiet". "Now you have to stay completely still". These cause them to yell or move a lot even at other times under the same circumstances.

10. If the youth is distressed, find out why or ask and, in this case, name the behaviour together with the possibility of preventing it

Example:

- M. (LN) is distressed because he has to get into a small taxi, where he has already hit his foot against something poking out and before he gets in he's distressed: "Don't worry, I brought a cushion and you won't hurt your foot".

- Others: "Nobody cares even if you yell, we have told them about your disorder", "We'll be able to get the X-ray done with no problems at all because there'll be two of us holding your foot"

11. Just because you don't yell at the boys doesn't mean you can't stop them, verbally as well.

Sometimes, precisely because these children can't control their emotions that are triggered by external factors, the caregiver's must act decisively, possibly accompanying their words with the movements to set aside what happened.

Example:

- M. (LN) wakes up during the night and can't stop himself from calling out continuously. His father gets up and says firmly "That's enough now, go back to sleep, I'm not getting up again" M. goes back to sleep

- M (LN) keeps crying because a request he made wasn't understood and acknowledged; his mother says firmly "Stop crying now because I don't understand; then try and tell me what you want again". M. calms down and tries to ask his question again.

Like all the earlier observations, whoever acknowledges and shares the suggestions for managing LN behaviour need to be very discerning with regard to how much the LN boy can do at that specific time. Too many expectations, as well as too much lack of confidence don't help get through moments of crisis "You can do it" can be as much of a trigger as "You can't do it"; the discriminating factor is the successful experiences that strengthen the possibility of success. Accompanying LN boys towards positive experiences, as stated in point 4, is delicate and personal. The earlier it takes place the easier everyday life becomes.

12. Spread the word

It is important for these suggestions to be understood and disseminated in all the areas of life and with all healthcare and operators who have anything to do with LN people

....the list can and must grow with the help of everyone who tries it out!