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## **LIFE EXPERIENCES: A BOY WITH LESCH-NYHAN SYNDROME AT SCHOOL.**

Michele is a boy affected by LND. Its renal failure was so serious that when he was seven, he was submitted to dialysis and when he was nine he had renal transplantation. He is fourteen years old now and he is finishing Primary school.

Michele came to our city when he was eight months and to our Motor Education Service when he was ten months. As psychologist of the service and expert in AAC, I approximately have known him since when he was two, because her mother had the feeling that "he could understand" more than what he showed to understand. She was right, but Michele's communicative initiative was not enough and in most part he wasn't understood by other people. With his smile and his look Michele held his relation with the world and he enjoyed the loving attentions of his big family. We begun a CAA project with his parents' agreement: we fixed YES and NO, I and YOU, the contextual indication, some gestures, some words and we built communicative tables that grew in complexity with photos and graphical symbols.

I quote from the book of Michele's mum a comment on that period: "... I remember the emotion I felt the first time that Michele put his finger onto the symbols on his 'table': He was still only expressing himself in monosyllables and it seemed that he could understand and wished to say more than we were able to decipher... After several meetings our boy began to try and express not only his discomfort and his more urgent necessities, but also his feelings, his emotions, his thoughts. How many pleasant surprises we had in those meetings: discovering that even if he seemed to be... a very small boy, Michele was growing. His curious mind was opening up to the world... The table now has been put aside because our young boy has learnt to say many words and is learning to write: but it has been a fundamental spring-board for him and us; a bridge on which to meet, suspended between his and our way of communication". Paola Mazzuchi Cargioli, *An (Im)Possible Life*, 1997.

The intervention was combined parent-child and the siblings were often involved, too. At the age of three Michele started to feel the impulse to hurt himself in a clearer and more intense way. The first modalities were arching of his back and neck and head banging, then he started biting his hands. One of the first words Michele said was "CO" when he asked the splints for his arms (the Italian word is "*manicotti*"), that prevented him to put his hands into his mouth.

All of us were quite worried when Michele started to go to nursery school. We and the parents decided to prepare the school by giving the little information we had on this rare and amazing disease, and we started a difficult adventure, with an attitude of search, each one with his/her own humanity, resources, limits, and role.

The second year of nursery school was terrible, we all felt a deep sense of dejection and impotence even after having found a system to avoid Michele biting his tongue and lips through a dummy tied to his hat. With a bitter consciousness that love was not enough, we wondered: What 's still going to happen? For a long time his mother had been asking me how to meet other parents, but then it seemed to me very difficult, nearly impossible. At the beginning of 1995, thanks to CNR in Genoa, I tried to ask for help through Internet: in three days we received fourteen answers. We quickly translated two articles written by Prof. Anderson. They were built on the base of a questionnaire to sixty families and we discovered that things were different from what was described on LND in the Children Neuropsychiatry handbooks as to the self-injury modes, the intellectual and social perspectives and life expectations. Information and contacts let us feel less lonely, we

acquired more confidence and this reflected in the work at school. In fact Michele began to enlarge the number of spoken words. Vocal language, natural modalities and augmentative strategies were integrated and we began to work on literacy, at first through the discovery of the permanence of the message for communication at a distance (tickets, postcards, invitations...) then by building texts in group of children, using the graphical symbols and an alphabetical code, they could read many times, show and give other people.

Since the age of seven the communicative table hasn't been necessary anymore: the oral language, even though sometimes is difficult to understand (Michele went on keeping his dummy in his mouth) became a quicker and more comfortable communication means. In the meantime Michele's family began to build the group of the families and to strengthen the international exchanges. Michele met other LN boys, and at the beginning he was a little frightened. I spoke a lot with his mother about the importance for Michele to know his disease, fundamental element of his "self" and of his experience of being in the world. Now in the moments of greater deject Michele vents his anger on a muppet called Nyhan, when Michele meets another child on a wheelchair asks if he is affected by "Nyhan" too....

With more precise information and ideas we organized meetings with all the staff and the parents of the elementary school Michele started to attend. He attended a normal class, in a particularly equipped school because it had a side for the most serious children (in Genoa we call them *Pollo*). In fact there weren't architectural features that denied the access to handicapped people and there were teaching and auxiliary staff having experience with disabled children, places where it was possible to carry an individual work or to go when Michele had crisis, remedial teachers, a larger auxiliary staff, the possibility of a reduced timetable (Michele has always had lunch at home). For the first time it was not the family to bring him to school, but a caregivers' cooperative. This involved other people to be informed on precautions and the sagacities that concur the well-being of Michele and on the possible arise of unexpected periods of discomfort and crisis.

The Town Council has not only intervened for the transport but it guaranteed the necessary devices for the positioning, the change and the personal hygiene and for the access to the learnings. In fact since Michele couldn't hold a pen or a book in his hand, the only way to access to the learnings forms is through a PC with a special keyboard that was chosen by our rehabilitation service and bought by the Town Council with the "Study Right" funds. Obviously things were not simple because everything could be a chance to hurt himself, his seat had to be padded with foam rubber, working time was short, Michele did not tolerate engagements and constrictions.... Michele played PC games alone when he was at home and he was learning to use the arrows on its expanded keyboard instead of the mouse, but he had to be on a good day to copy or write a word down as a dictation. We started to understand that the reduction and the control of the stressful events are the conditions of Michele's well-being, and his well-being is the goal and the premise of every plan. But then, both for parents and teachers, how recovering and modulating the educational function? Which are the ways "to be with" that help him to go ahead, to deal also with homework and learning?

For all the 5 years at Elementary School Michele hasn't had a good attitude towards Maths, he didn't like it. He preferred listening and he achieved a good competence in comprehension, even about subjects far from his every-day life: history, geography, and above all science. At the end of the fifth class the way towards literacy acquisition was still long. However there was a thing we had understood all together (parents, therapists, teachers): the best way to obtain motivation, interest and participation towards literacy in Michele was to use functional language (tell, write a card, an invitation...) and linguistic games. But more in general humour, thoughtlessness and irony were Michele's padding of life.

Michele started "Scuola Media" and because of his frequent non-attendances at school,

due to fevers, day hospitals etc., a project of a two-year third class was thought for him. (Michele's mother often reminds that Michele is not only disabled, but also ill). The school Michele attends is the public school in the neighbourhood.

Here is in short an interview with Michele's specialized remedial teacher, Prof. Bruna Danello:

*What have you learned in the last three years with Michele, that can be helpful for others?*

According to my experience with Michele, I think it is important:

- not to minimize, but take it seriously when he feels ill
- to know the reasons that give him anxiety very well and find strategies to reduce it
- to use irony in order to both reduce anxiety causes and make didactics easier
- to use non verbal communication (look, mimics, expressiveness) both spontaneously and consciously

In general it is important to start from him, to work in reply to what Michele says. His contents are mostly related to school environment, hospital and family. Only Michele, however, can joke on his family, because if other people do it, he feels offended!

*Can you give me some examples, please?*

I have learned to tell him recurrent witticisms. For example: Michele comes and asks me "How are you?" and I reply: "I'm sweetly getting old" or "I stand on my tomb".

When he feels ill we usually go to the entrance hall of the school where there is a poster of the Archangel Michael: I must punch his patron angel because he sleeps instead of protecting him. Michele then smiles. Now he sometimes says that "It's not fair", speaking about his disease and I sincerely tell him he's right.

Then I use a kind of complicity. I tell him: "Today Dr. Alessandra is coming" and I know that he gets excited, then I tell him to think about lies to tell the doctor, something wrong for example 'the lift never breaks down'. Or I use slightly unconventional words and we tell them to the Italian teacher who is passing by.

I make use of strange themes as a teacher: if Michele thinks to suffer a wrong, he often cries, then I take his attention by telling him: "Do you want to go and see the beautiful girl in 3 F? Or the nice girl in 2 C?" They really are the most beautiful girls in the school and we go and see if they're passing by.

If we play a didactic game like "hangman" with a classmate and this makes a low score we make fun of him. Michele must win every time, he can't stand losing. When once his class played a volleyball match, we supported his classmates by waving an Italian flag. His class was losing and he burst into tears.

Sometimes I use a "hard" irony: I say aloud: "This is not a cinema. This is a school!".

Michele worries a little, then the specialized operator mediates. This is a very important strategy, a three-player strategy. We often make irony together: we observe and comment other people, in class, for example, we hear the teacher of Italian saying for the umpteenth time. "I don't want to get angry", I ask him: "Will she succeed?" He laughs and says "No".

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CA RO SI N DA CO  
MA S SI MO NON C'E'  
NON E' AL LA VO RO  
ED IO SO NO SO LO

*Which are the principal causes of anxiety and worry at school? How can they be faced?*

In the first class noise was a source of anxiety, both shrill and low sounds when they were sudden or excessively intense. This fear has now reduced. A great source of anxiety is the absence of an adult. It is clear that Michele feels safe when there are two grown up people, Michele needs two people. Generally it's me and the caregiver, in Genoa we call them O.S.E. We do not stay necessarily together, but we must be present. In the first year there was a young woman, Marina, since the second class it has been a young man, Massimo. Sometimes Marina was late, so we used to make fun of her. When the actual caregiver is late I tell Michele: "Perhaps a blond girl substitute is coming", in order to reduce the anxiety he feels when he thinks to be left alone. Having a second help, who is not a teacher, is a very important thing for many reasons: to relieve the didactic engagement and sometimes to give a hand the teacher, because it is difficult always to be at the fore front; being three people gives us the opportunity of keeping a good equilibrium. Sometimes it's Michele himself who tells me: "Go and smoke a cigarette!".

Another source of anxiety is when, during the break, his male classmates play fighting. We have managed to reduce it by letting him participate or organizing the games better (bumper cars, pursuits...).

When Michele gets angry, he tells me: "I send Marco to you". Marco is Michele's very tall and strong brother that he calls to "hit" people when they need to be punished. Sometimes I tell Michele: "No, I don't allow you to go home, because you will send Marco to hit me."

When he is really worried because he is going to hospital, we make "war plans": for example we plan to go and burn Gaslini Hospital with my cigarette, or we send my dog to bite the doctors. The plan can be articulate: firstly we send Massimo to put some bones inside the Hospital, then we send my dog that then bites. We play as we were a gang.

Planning to take flight is a good way to "channel" his anger through game. For example we leave for a foreign country we studied in geography by car without his parents. While flying we run into all people we don't like.

Another thing that seems to help him is thinking about threats of surreal punishments, for example "For Christmas we'll give you a rubber snake as present".

Sometimes Michele swears at me, he calls me "old mummy", or "skinny". In these cases it is important neither to ignore, nor to reply, but stop for a while and smile consciously, sometimes he excuses himself by saying it is "Nyhan". Sometimes he can control himself and then he feels very satisfied, because he can say "I succeeded"

Sometimes he feels strange fears, for example of being taken ill or of bad weather (in his

opinion bad weather worsens his self-injury). Also in this case I found a way to reduce this fear: once it was going to rain and we saw a lady who was hanging some clothes out, since then we have been going to see that “stupid woman who had hanged the washing out”.

These things seem to work, but I must say that they don't work every time and it is becoming more and more difficult.

*Can you tell us how you apply all this to didactics and in particular to literacy didactics?*

We start from some examples whose goals are to waken his interest on what is written and let him understand the power of written language. Once during music lesson the teacher about the theatres in Genoa. I had Michele's copybook and the felt-tip pens in front of me and I told him “In Genoa there is *Genovese Theatre*” and Michele commented “No, *Doriano*”<sup>1</sup>. So I wrote “*Doriano Theatre*” in capital letters on his copybook and he was really attentive to check that his joke was correctly written.

Another time during geography lesson, the teacher was explaining Padana Plain and I repeated “Padana Plain” and Michele commented “Bossi”<sup>2</sup>. So I wrote “Padana Plain – Bossi” on his copybook.

The activity on the names started by chance. In that period I felt that Michele needed to work on the theme “death”. He used to joke by telling me “Bruna is stone dead”, but I understood that it was not only a joke. I drew a lot of tombs on the blackboard (obviously not of his family) and Michele dictated the names to write on them: Luigi, the headmaster, the classmates... The game finished when the blackboard was full of tombs and Michele told me “I'm scared” and I replied “All of us”. Now when we speak about tombs, we can smile, and we can also draw Heaven: it is out of doubt that in Heaven at least Michele wants to be able to walk.

In order to let Michele build sentences even orally and to work on the pronouns I/YOU, I told him: “You think and I write”. Now Michele builds sentences.

Something he enjoys himself doing is looking for the writings on the walls of the school. We found “Burn it” (they obviously intended the school). Even this is a way to learn reading!

He enjoys himself writing marks and announcements. For example:

“A black mark: Bruna has offended Michele”

“Announcement: no homework, only games”

“To the Mayor: Massimo doesn't let Michele play”.

*Please, can you explain to us how you organize an activity?*

Last year we used Linus strips to improve reading and I obviously chose ironic strips. They usually are divided into four images and the written text is not too long. To work on writing we use PC. Michele identifies himself with Snoopy and through Snoopy he speaks about himself. The description of the cartoon is not interesting to him. He is interested in having a chance to speak about himself. For example: “Dad is playing fighting with me”. We always work on his own experience. Snoopy's day is Michele's day.

When I ask him to write it is very important to reduce his motor task, so I usually write the recurrent word, such as the name “Snoopy”. The result can be:

SNOOPY BITES THE BOY

THE BOY IS SCARED

THE GRASS IS RED (dirty of blood)

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1 In Genoa there are two football teams: Genoa and Sampdoria. Michele is a supporter of Sampdoria

2 Umberto Bossi is an Italian politician who proposes the secession of the northern regions from the other ones to create a new state called “Padania”.

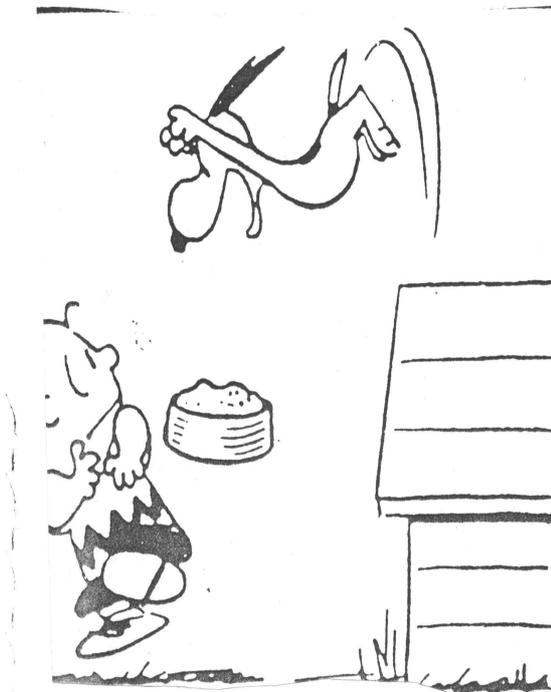
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**COSA FA?  
SNOOPY SALTA SUL BAMBINO PER  
MORDERE.**

**LINUS HA PAURA.**

**SNOOPY MORDE**

**L'ERBA E' ROSSA DI SANGUE**



A new and very interesting activity we chose this year was to read pictures drawn from books or the Internet. We made a list of 6 kind of feelings: beauty, joy, sadness, restlessness, serenity, anger, as they are useful to talk about those pictures. I chose paintings by Renoir, Chagall, ...Michele is impressed by such details as the purple hovel, or he sees Santa Claus as part of the picture. Maybe I was wrong in showing him "The Howl" by Munch: he only said: "He's an alien".



Then I showed him Goya's "Dressed Maya" and I asked him what the nicest detail was to him. Without hesitating, he simply answered: "her legs". Van Gogh's painting showed the sea by night and Michele, who saw a very similar landscape with his mum, in Genoa, was really touched.

He's bound to his mother by great affection. One day he burst into tears because I was not able to understand his dream: he was a sea captain and he was married to his mother, but then he was overwhelmed by a grievous sense of lost.

*What do you remember of the day you first met Michele?*

The first year was very hard. He could cry so much that at 8.20 he was home again. Some teachers were scared. It lasted two weeks. When I arrived I was told nothing but he had motor problems. When I first saw him I thought "What beautiful eyes, they look ironic!".

I think this interview is extraordinary, because something so complex and emotionally touching is told in a very clear and simple way. I think we could dwell upon many different aspects. As a psychologist, I can't help naming just two of them. First of all, it is basic to create a link with Michele, a real contact; he is very sensitive to the others, he can feel whether you are sincere or not. Secondly, learning to control the strongest emotions (particularly fear and aggressiveness) is also very important, by sharing his own feelings and through irony (which is game and communication at the same time). These are basic emotions, as well as the more positive ones, but we have to learn how to cope with them, and it is a real gift; this way, we can help Michele in finding his own balance. (For instance, in Munch's Howl such definite restlessness should be driven away as alien). As a matter of fact, Michele needs a character/scenography he can identify himself with, his desires and experiences, by creating situations where to direct his aggressiveness. Some subjects also seem to allow him to face the thought of death and the fear of both destroying and being destroyed. The theme of biting is basic (the dog biting, Snoopy biting the child). Today sexual aspects are also likely to emerge, which should be faced.

It is very important to study more closely life experiences of people affected by Lesch-Nyhan Syndrome, Michele's everyday life can teach so much to everyone, as well as other people's everyday life can be helpful to him.

Even when Michele feels fine, it is useful to help him relive his dramatic life condition, which is due to the strong physical-emotional relationship, so peculiar to him (I cannot help imagining the two things as linked to each other), as well as to the fact that he cannot easily give vent to his own feelings, through motion.

I'd still like to talk about Michele's school experience. He has always attended public schools and he regularly started school at the age of 3. It demanded great energy to the people who said: "yes, I will try to do my best". I esteem and thank the remedial teachers who helped Michele: Marina, at the infant school, Carola, who already had got her specialization, Rosa, who chose her experience with Michele as a subject for her graduation thesis, Caterina, who was so scared at the beginning, but then learned to laugh with Michele, and then, at the secondary school, Bruna, whose interview we could listen to. I believe that when a teacher first meets Michele, he/she instinctively reacts with fear and a sense of inadequacy, and it is logic; we should worry if it wasn't like that. But I strongly believe that isolation should not be allowed. It is very hard to learn to set aside your teaching goals to continue only once Michele's physical conditions allow you to, to learn to defer all your projects, expectations, to share the pain sometimes, while keeping hope and faith alive, living day by day. I hope, as a psychologist who works for the civil service, I have proved myself able to support, understand and share everything with the school staff I've been working for.

The choice of letting children affected by Lesch-Nyhan Syndrome attend public schools rather than special schools (which usually upsets foreign experts) is very good, from different points of view: on one hand, Michele's schoolmates never tried to imitate his behaviour (and this greatly worried many foreign schools), on the contrary, they tried to help him to restrain his behaviour such as spitting and swearing, which he calls "Nyhan" and apologizes for that); on the other hand, it was possible to use irony and language game, which demand great mental flexibility; besides, such spurs as writings on the wall we can find in a school could not be artificially reproduced.

Getting to know and learning to live with children who have the same problems and limits as they have is not real alternative to attend a normal school; they can be both good things. However it is also true that such things are possible under certain circumstances and when facilities exist; I'll give some examples: lack of architectural features that denies access to the handicapped, classrooms where to stay for individual activities and when Michele feels bad, prevention of risks for self-injurious behaviour, special devices (new technologies are basic), timely and correct information about the main features of the Syndrome, cooperation among the teachers, involving not only teachers but other adults, presence of specialized caregivers as the Law foresees, everyday school-family communication strategies, also guaranteeing urgent communication. Generally, family-rehabilitation therapists – school cooperation is basic for planning and managing school attendance.