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**Telling stories, telling lies:
The importance of a progressive full disclosure of DSD**

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..... healthy mental growth seems to depend on truth as the living organism depends on food. If it is lacking or deficient the personality deteriorates....

W.R.Bion (1965)

James Pennebaker (1996, 1997) wrote that the possibility to speak about traumatic experiences is an important healing factor. On the contrary a secretive attitude about aspects of one's personal history contributes to many forms of behavioural inhibition and to a major incidence of illness.

He maintained that putting painful experiences into words enables people to organize and integrate feelings and thoughts and it gives the feeling of being able to foretell and master their life. If an experience has a narrative structure and a meaning, it is easier for the people involved to cope with the emotions and the memories, even bodily memories linked to them.

Karen

The first person with DSD I met many years ago, was a fifteen year old girl with 5-alpha-reductase deficiency, who thought she came to the hospital because of her obesity.

She didn't know very much about herself even though she was sent to a psychologist who was asked to "tell her everything", that meant she could not have periods, and she could not have children.

She didn't know what the problem was; she had only been told it was something to do with "some ovarian glands that didn't work and for that reason the ovaries had been removed".

Merely remembering her sessions with the psychologist made her burst into tears, she didn't want to see her again. She underwent gonadectomy and vaginoplasty but she didn't even think of doing the vaginal dilatations she was advised to do. She acted as if the lower part of her body didn't belong to her anymore, she didn't get undressed even to have a bath and she agreed to be examined only under general anaesthesia.

The story I was told in a condition of overwhelming anxiety was a mass of incoherent fragments of missing truths, half truths and lies where a psychological intervention looked like the surgical operations the girl underwent without any preparation or participation.

The results both in psychological and in medical terms looked awful so I felt the need to know more. I was curious to know more about Karen but I left her the choice to decide whether to have other sessions or not. She didn't want to and I respected her choice. I only spoke with her mother when she contacted me -with Karen's permission- and after some years I was told that Karen showed her mother an internet site saying without any particular feeling : "Look! 5- α -reductase deficiency, that's what I have".

If she took possession of her history without any particular reaction what would have happened if she had been allowed to know the single elements of her personal history and the meaning , if there was one, of the operations she underwent ?

Rosa

Rosa was sixteen years old when she was diagnosed with complete androgen insensitivity. A gynaecologist consulted for primary amenorrhea proposed a chromosomal test that found an XY caryotype.

She was not fully informed about her condition and when she underwent gonadectomy due to a presumed tumoral risk, she was told her ovaries had to be removed.

Six months later I met her, a smart and clever teenager who attended a classic high school with very good results. She told me that she had willingly submitted to clinical tests but she still couldn't stand physical examinations and she was still nervous about coming to the hospital.

She felt as if she was being raped because a decision about her and her body was made without asking her. Her grief was greater because she thought that she could have shared in making the decision about the surgical operation.

How will she feel when she realizes that many other things were kept from her?

These are only two of the many stories all of us know very well and we cannot console ourselves thinking they are the fruit of old practices.

The old way of thinking was that to reach a stable gender identity it was important to have unambiguous genitalia and to be raised without ambiguity in the chosen gender. Early genital surgery was considered necessary as if any bodily sign of ambiguity was considered a "foreign body" that should be removed as soon as possible. In accordance with that there was a consequent withholding of the diagnosis as they thought that a full communication of DSD would have a traumatic effect and it should be avoided.

The word trauma can be used to refer to the psychic impact of an event, a separation, a pain, an illness exceeding the subject resilience and her/his capacity to deal with it and to elaborate it in a psychic way. Trauma gives rise to the experience of helplessness and massive deconstruction when the ego and its functions are overwhelmed by a degree of pervasive anxiety that is impossible to master.

Freud (1893) in his first writings used the metaphor of the “foreign body” to describe the action of trauma “*the trauma or more accurately, the memory of trauma acts as a foreign body*” for the psychic apparatus. A memory unacceptable for the conscience should be removed, but once repressed, memories continue to have an influence and if not faced may give rise to an anxiety that floods the ego and opens the door to the realm of psychopathology.

In DSD the newborn body is often felt by parents and health care workers as a foreign body, different from anything that was expected. It rouses anxieties, concern and alarm that create in us a foreign body reaction that make us try to expel it.

But we often forget that what for an outsider metaphorically is a foreign body, is the body a person is going to live with for the rest of his or her life. The body and its sensations are the original matrix of any later emotional experience. Acceptance allows us to experience the sense of physical and emotional continuity, which are the basis of our personal idiom, the skeleton of the story of everyone’s life.

One of the fallacies in the practice of withholding is that children don’t remain children forever and sooner or later the truth will come out, frequently all of a sudden and without any preparation as happened in Cleo’s story that is quite similar to many others.

Cleo

Cleo was 26 years old when she went to a new gynaecologist because of a change of town when she moved to a new job after her graduation. They soon understood that she had an XY chromosome and probably she had a partial or complete androgen insensitivity. That discovery, all of a sudden, shocked her. As she went out of the Clinic, in a burst of rage she entered the first telephone box she found and called home asking her parents if they knew and if so, with increasing anger, why they didn’t tell her anything. She was 26, with a degree and she was living by herself a thousand miles from home. Wasn’t it right to know what was so important for her, what she was? She slammed down the telephone and in that moment she imagined that the four glass walls of the telephone box were windows open on her new life. For a long time she refused to see her parents and even though she was able to forgive them, the relationship between them remained impaired. Little by little she started anew putting together the pieces of her stolen history and the new one even though it was full of anger and regret, was full of awareness and of other feelings, but most of all, it was a true one, it was her story.

It is now widely accepted that one of the most painful issues related to the DSD condition is the lack of information and the feeling of secrecy regarding the clinical history of the subjects (Slijper et al. 2000, Wisniewski. et al. 2000, Creighton S., Minto C.L. 2001, Carmichael P., Ransley P.G. 2002, Brinkmann L. et al. 2006).

I was privileged to be invited to some meetings of AISIA, [Associazione Italiana Sindrome Insensibilità Androgeni] the Italian Association of persons with AIS and

there I had the confirmation that one of the most frequent regrets is not having been informed about their condition by the people they trusted most, parents, physicians, or parents and physicians together.

On the contrary, the possibility to speak in a free, open, sincere and friendly way about their condition is a way to work through aspects of their stories that before were sources of pain.

With AISIA we started a study on DSD and the first results seem to confirm that the full disclosure is usually between 16 and 26 years of age and nearly always all of a sudden, after years of lies, consulting gynaecologists, surfing Association websites, eavesdropping on parents and medical staff talks.

I don't think there is a way to communicate a severe diagnosis without pain, but the fear of the traumatic impact of an experience is often due to a lack of distinction between pain and anxiety.

Pain may be the cost of psychic integration; non integration is a source of anxiety and anxiety is the impossibility to feel true pain, with its intrinsic transformative potential.

Suzy

Suzy knew about her conditions when she was sixteen years old and went with her parents to the gynaecologist due to primary amenorrhea. She and her parents together were told very sincerely her diagnosis and what that would imply.

“I experienced a great pain and sometimes I feel it now, but like all painful experiences it contributed to strengthen my character in a positive sense. It has matured me quite a lot and made me understand what makes life worth living. Sometimes I feel a little sad but my basic character hasn't changed.”

Two people

Like Suzy's, the reactions of the following two people who described their moment of disclosure, were not too traumatic.

“After I found out the truth I had a feeling of peace because I found an answer to the many doubts and to the many anxieties that I had had since I was a small girl and I could not find a reason for all those examinations, those therapies, the lack of menses, the physician's comments and all the rest.”

“I felt a grief that left behind a veil of sorrow and of awareness. I felt as if I had lost my identity and I felt disappointed that I had not been informed before. All those feelings were softened by being able to talk about my condition and share it with someone.”

The traumatic psychic experience doesn't usually appear out of the blue. It usually appears with a double origin, when a fact, a memory which when it happened didn't have a traumatic impact may be traumatic due to the increasing maturity of the child. Especially if what happened was kept secret beyond the possibility of the subject to know it, to find a reason or an explanation for it.

The body, as well as the mind has its experiences, its pains and its memories. Early operations with their long and inevitable presurgical and postsurgical phases should be limited to those strictly necessary. Sensorial memories lie in our senses and require a reason that can explain what happened. The impossibility to transform them into mental images, into thoughts, into words is what transforms a psychic or a bodily pain into the first moment of a traumatic experience.

If that is not possible, a pain cloud hovers over what happened and was kept away and may constitute a potential traumatic break that may appear again at a later time.

I think that if the truth is not told when the opportunity is right, when the problems are on the table and the situation is emotionally warm, it is very difficult to tell the truth in the future. The thought that the future will be a better moment is the first step in a sequence of omissions that will never find the right time to be interrupted.

Children cannot be kept away from the pain and the losses of life, it is important that in those inevitable moments they are allowed to cry but the most important thing is that in those moments they find someone who can comfort them and share their tears.

It depends very much on the way the person is helped to cope as was pointed out by Ferenczi (1934) who considered that lying and deceitful information could be a potential traumatic factor "*the adult behaviour toward the child who suffers from a trauma is part of the psychic action of the trauma itself*"¹.

Balint (1969) developed this concept in his three phase theory of trauma.

The first phase sees the immature child dependent on the adult and although frustrations in their relationship may occur and lead to irritation and even to rage at times, the relationship between the child and the adult is mainly of trust.

In the second phase the adult, contrary to the child's expectations, does something very arousing, frightening or painful; this may happen once and quite suddenly or repeatedly. It is not necessary that the adult actually does something, it is sufficient that something, an illness, an accident occurs in the framework of the relationship

The real completion of trauma lies in the third phase when the child tries to get some understanding, recognition and comfort and the adult, either because he also is in pain and traumatised or he is trying to avoid greater suffering for his child, acts as if nothing had happened.

Here we have a further confirmation that in these situations the parent's pain must necessarily be taken into consideration; it must be held and contained to prevent its becoming a pain that is always alive, full of indigestible emotions.

The parent's role is of primary importance because it's up to mummy and daddy to find and give a sense to the experience the family is living. The discovery that the son or the daughter has something that is different from what was expected may have a great influence on the relationship to come. Only the progressive acceptance and the

¹ Ferenczi, (1934), Opere Complete, vol. IV, pag. 103.

recognition of the real him or her, the real boy or the real girl, may avoid that at a more or less conscious level some sort of refusal will invade the relationship.

No kind of DSD is easy to deal with or to think of. We have no ready made solutions. The presence of DSD reactivates fantasies, fears, conflicts so we must recognise, that, as happens for parents, we are, at least temporarily, traumatized and pushed to defend ourselves. This is the main reason why old practices die hard.

A way to protect ourselves from the trauma is to assume active aptitudes while it is important to respect the timing and the feeling of the persons and not to present ourselves as omnipotent experts. We now know that these issues cannot be dealt with alone. The importance of real team work is unanimously accepted where opinions can be compared; anxiety can then be held in check. Group work can create room for thinking and help parents to decide giving them all the pros and cons of the situation.

We try to reach the best professional levels in medical, as well as in psychological practice, we are often pushed to act, while in this field we are often required not to do so, not to give answers and prescriptions. Our technical skills are challenged by what Bion, using the word of John Keats, called a "*Negative Capability, that is, when a man is capable of being in uncertainties, mysteries, doubts, without any irritable reaching after fact and reason.*"(1970)

Most of my contributions are taking steps back, I am at people's disposition if they need to speak to me, but mostly I listen, people with DSD need to be listened to, especially by their physicians and their parents.

A good way to inform patients about their condition is the progressive three steps suggested by Sljiper et al. (2000).

The first step regards early good information on the biological and psychological aspects of the normal sexual development, because "it is too difficult for a child to comprehend the deviation in the development of its genitalia when it is not informed about normal development".

The second phase that should start around the age of 11, before the starting of possible substitutive therapy, should contain all the information except that regarding the XY caryotype. Full information should be available at the age of 16, 17 and that would be the third step.

I fully agree with that but I would stress the importance of the first step also for a different reason. A good sexual education is very important for children not only for the knowledge it provides that till preadolescence is very relative. Mostly because it can give words to name curiosities, discoveries, fantasies about the body and its drives. But it is even more important because it enables them to know, tells them in an implicit way that no knowledge is hidden or forbidden and that the adults support their right to know and that they can trust their parents.

When things go sufficiently well, children know that there are words to give a name to their confused bodily sensations, that they may build their own story about the mysterious world of the body even though the real meaning of their knowledge is not yet completely mastered. Names are not important for the objective reality of what

they are for, but because their use means knowledge, to explore the inner and the outer world.

Only maturity will allow children to acquire the full comprehension of what a name is for, but if names are told, the children's need to know will not be hurt by realising that they weren't considered capable of understanding and they were told lies.

I also think that even the negative impact of the XY problem may sometimes be overestimated.

Sally

According to her mother, a decent person, apparently sensitive and emotionally close to her fifteen year old CAIS daughter, Sally had some knowledge about her clinical condition, about not having uterus and ovaries, about her infertility. She doesn't only know about the Y chromosome because at the moment her parents think she is too young to know.

They learnt about Sally's AIS when they went to the hospital for a suspected inguinal hernia when Sally was two months old and the surgeon turning to the mother said "but madam that's a male!".

The parents little by little apparently recovered from this shock but they wanted to spare their daughter. They see Sally as an even tempered girl with many friends, clever at school and able to cope with the burden implied by her condition.

She underwent gonadectomy when she was three years old, when she was six she had vaginoplasty, and now she is undergoing substitutive hormonal therapy. At the moment Sally comes to the hospital twice a year for an endocrinological check up.

I left the mother with the impression of a quite good situation where a careful and genuine parental attitude promoted a good integration of Sally's situation. Meanwhile a colleague had a meeting with Sally.

Sally is a tall, charming, attractive young woman. At the beginning she is reluctant to talk to a shrink but at the end she speaks for one hour and a half alternating talking about herself, letting out emotions and crying.

Her regret is mainly about not having anyone to talk to about her situation, she feels she cannot speak to her mother but feels the need to talk to someone, the best would be a person who had passed through what she is passing through now.

After a long silence she starts crying thinking she will never have menses or children. She has been coming to the hospital since she was a baby and only a couple of years ago her mother told her the reason why she comes and why she takes pills. She thinks that if she had been told earlier probably she would not have understood. When her mother told her the pros and cons of her condition, Sally started to cry even though she is still not sure she has understood everything.

She doesn't speak to her brothers because she is afraid they will let something slip with friends. She has never had the opportunity to speak about her condition with her father, although he knows everything. She doesn't speak with friends because she fears

that someone could use “this thing” to damage her. She doesn’t like to talk to her mother because she realizes, despite her mother’s efforts not to show it, how badly she suffers and always suffered, even before Sally discovered what for. Anyhow Sally thinks her mother is not aware how much she suffers for her problem because she conceals it from her.

She knows she underwent four operations that she doesn’t remember. She knows she was two months old when the operations started to remove an inguinal hernia and soon it turned out she had no ovaries.

She senses that this is not the whole truth, that there is much more to know, she fears it is something about cancer. For the moment she feels she is not able to ask more “perhaps I will understand better when I am eighteen years old and I have studied what an inguinal hernia is”.

Sally shows us how complex the situation is and I am not sure that the XY problem would have been worse if it had been told with a more open communication.

As I said before, the whole thing is not easy, solutions are easier said than done. I think what we can honestly do, little by little, walking step by step with our patients, is to give the people who ask for our help, parents and patients, all the elements they need to build their personal story even though they seem to us to be too painful, too frightening to deal with.

Because, at the end of the day, the old practice showed its limits, and we should learn from our mistakes. Those are their stories and even though we can play an important role in them the basic thing is that it seems to me that we have collected enough evidence to show that our patients ask us to share with them the difficult but rewarding path towards open communication.

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