Responses to falling III: defense mechanisms used by women with Turner syndrome and variants

Respostas ao adoecimento: mecanismos de defesa utilizados por mulheres com síndrome de Turner e variantes

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Abstract

Background: Article extracted from the doctoral research entitled "Experiences of the infertility phenomenon by patients suffering from Turner syndrome and variants: a clinical-qualitative study" approved by the University of Campinas. **Objectives**: To understand the defenses employed by women suffering from TS and different ways of dealing with the disease. **Method:** Qualitative research with exploratory design, non-experimental. The instrument consisted of a semidirect psychological interview, involving 13 women, undergoing semestral medical follow-up at the Center for Integral Attention to Women's Health (CAISM), and whose sampling was determined by saturation. Data was interpreted using the psychodynamic approach along with an eclectic framework of theoretical references for discussion in the spirit of interdisciplinary approach. **Results:** These women displayed psychosocial conflicts such as difficulties in interpersonal relationships; feelings of resignation, anger, impotence, devaluation and depression symptoms. Defenses used were: repression, denial, annulment, fantasizing, adaptation and sublimation. **Discussion:** Women suffering from TS and variants must deal with the disease's organic and psychic implications that cause great suffering and often hinder a saner social insertion. In this case, the study's findings can guide ambulatory psychological support concomitantly to the routine clinical protocol.

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Key-words: Turner syndrome, defense mechanisms, qualitative research, genetics.

Resumo

Objetivo: Conhecer as defesas utilizadas por mulheres com síndrome de Turner (ST) ou formas variantes para lidar com a doença. **Método:** Pesquisa qualitativa com desenho exploratório, não experimental. O instrumento consistiu em entrevista psicológica semidirigida, aplicada em 13 mulheres, cuja amostragem deu-se por saturação, as quais fazem acompanhamento semestral no Centro de Atendimento Integral à Saúde da Mulher. Os dados foram interpretados utilizando-se da abordagem psicodinâmica, aliada a um quadro eclético de referenciais teóricos para discussão no espírito da interdisciplinaridade. **Resultados:** Essas mulheres apresentaram conflitos psicossociais como dificuldades de relacionamento interpessoal; sentimentos de resignação, raiva, impotência, desvalia e quadros de depressão. As defesas utilizadas foram: repressão, negação, anulação, fantasia, adaptação e sublimação. **Conclusões:** As mulheres com ST ou formas variantes têm de lidar com as intercorrências orgânicas e psíquicas da enfermidade provocando grande sofrimento que, frequentemente, dificultam uma inserção social mais sadia. Neste caso, os achados deste estudo poderão nortear acompanhamento psicológico ambulatorial concomitantemente ao protocolo clínico de rotina.

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Palavras-chave: Síndrome de Turner, mecanismos de defesa, pesquisa qualitativa, genética.

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Introduction

This article deals with the defense mechanisms used by women with Turner syndrome and variants so that they can psychologically structure themselves in order to face the illness's inter-currents. The article was extracted from a doctoral thesis research realized at the Center for Integral Attention to Woman's Health (CAISM) entitled: "Experiences of the infertility phenomenon by patients suffering from Turner syndrome and variants: a clinical-qualitative study". The research was approved by the Research Ethics Committee of the Faculty of Medical Sciences, University of Campinas (Unicamp) on the 16th of July 2002 (protocol Nº. 270/2002) and the thesis defended in February of 2005.

Ethical aspects

Each participant was informed of the research's objectives and procedures. Data was collected after two copies of the free and clarified consent form had been signed, one copy remaining with the interviewed patient.

Clinical characteristics of the syndrome

Turner syndrome and variants are associated to total or partial monosomy of the X chromosome, whose incidence reaches one in every 2,500 girls[1]. The characteristics of newborn carriers can be similar to those of normal children, except for their low weight and sometimes by the lymphedema of their extremities. To this end, the majority of diagnoses occur during early childhood because of the child's accentuated low height, or during puberty in the face of the absence of the secondary sexual characteristics development and primary or secondary amenorrhea.

At adult age these women can present the entire syndrome's phenotype spectrum, such as autoimmune thyroiditis and renal and cardiac abnormalities, depending upon the extension and location of the chromosome lesion. The hormonal profile is characteristic, by reason of ovarian insufficiency, bringing about low levels of estrogen and high levels of follicle-stimulating hormone (FSH) and luteinizing hormone (LH). Infertility shows itself in almost the absolute majority of cases, as an irreversible situation.

As variants, the genotype and phenotype amplitude can be understood to be characteristic of the syndrome. The total deletion of the X chromosome's short arm frequently results in the classic phenotype, whilst smaller alterations are associated with low height, primary or secondary amenorrhea and infertility. The deletions of the long arm denote a wide range of phenotypes, although only small portions are directly related to the syndrome, especially those linked to gonadal development and the ultrasound test will reveals diminished or streak gonads.

For treatment, the principal recommendation is hormonal reposition therapy (HRT), looking towards the prevention of cardiovascular illnesses and osteoporosis. At some study centers, for the younger patients, there can be the indication of using a growth hormone (GH) and of estrogen, as treatment for low height and the construction of a female biotype that is not infantile[2].

Defense mechanisms

This is a group of operations carried out by the ego in the face of the dangers that come from the id, superego and external reality. Defensive behaviors are not exclusively of the pathology and they normally contribute to the adjustment, adaptation and balancing of personality. Each person shows evolution in their defensive structures and that, which might have been adequate in childhood or in adolescence, may not serve in adulthood. Therefore, this defensive repertoire can enunciate plasticity or rigidity. However, frequently every individual subconsciously selects a number clearly restrictive defensive behaviors, which they make use of to deal with the conflicts coming from their internal or external world. That is to say, whatever defensive structure on being built up as a subconscious choice to operate pushes away other possibilities, which then produce a certain functional limitation of the ego[3-5].

Notwithstanding, all of the defensive mechanisms require the expenditure of psychic energy, some being more efficient than others. The mechanisms considered more mature are those that demand less energy expenditure, in the sense that they are successful and generate the stoppage of that which is rejected. While the mechanisms considered neurotic or immature, by being inefficient, demand a high expenditure of energy because of the repetition or disturbance of the rejection process[6,7].

Subjects and methods

In this cross transversal study the clinical-qualitative methodology was adopted, which involved an individualization and refinement of the qualitative methodology and is conceived as a scientific means of understanding and interpreting the psychological and psycho-social significances that the people linked to health problems attribute to phenomena in the field of health-illness. This methodology is supported by three pillars: **existentialist attitude** which appraises the anguish and anxiety present in the individual to be studied; **clinical attitude** involving leaning towards the person and listening to their emotional suffering with the desire of offering help: and **psychoanalytical attitude** involving the use of the conceptions coming from the subconscious dynamics of the studied person, for the construction and application of instruments to be used in the research, as

well as for the theoretical referential in the discussion of the results[8].

A psychological interview[9], semidirected with open-ended questions[10], was applied, as the instrument, to 13 women with phenotype compatible to TS and variants, involving semester or annual accompaniment at the CAISM-UNICAMP, during the period between 1990 and 2001; their ages varied from 18 to 45 years. The intentional sample[11], since those interviewed were contacted after verification at the Attendance Records Office, gave us the freedom to choose the subjects looking towards attending to the research's specific objectives, and was closed via the criteria of saturation[12]. That is to say, the interviews were ended from the moment at which they began to become repetitive, adding little of new substance and collected data being sufficient for an in-depth discussion. The recorded material, which was later transcribed, was submitted to Content Qualitative Analysis from which emerged as one of the categories, the defense mechanisms.

For the interpretation of the data, a psychodynamic approach was used, allied to an eclectic framework of theoretical references for the discussion in the spirit of interdisciplinary cooperation[13]. Validation was made in weekly meetings with the paired personnel[13], at the Laboratory of Clinical-Qualitative Research (LPCQ) of the Department of Medical Psychology and Psychiatry (FCM/Unicamp). For the identity preservation of the women who participated in the research, their names were substituted by names of flowers.

Results

Socio-demographic characteristics and the clinical situation at the moment of interview

Rose = 30 years, high school completed, housewife, married, height 1 m 45 cm, weight 67 kg, underdeveloped breasts and clitoris, discrete femoral osteopenia, karyotype 46,XX/ 45,X.

Violet = 31 years, second year of high school, nursing assistant, married, height 1 m 55 cm, weight 49 kg, underdeveloped breasts and uterus, discrete osteopenia in the vertebral column, karyotype 46,XX/ 45,X.

Jasmine = 31 years, university degree, teacher, single, height 1 m 63 cm, weight 65 kg, underdeveloped ovaries, discrete lumbar osteopenia, karyotype 46,XX/ 45,X.

Orchid = 22 years, high school completed, production assistant, married, height 1 m 42 cm, weight 45 kg, underdeveloped uterus, karyotype 45X/46X,iso X(q).

Daisy = 45 years, primary school completed, housewife, height 1 m 49 cm, weight 73 kg, underdeveloped breasts, uterus and ovaries, osteoporosis of the lumbar column, karyotype 46,XX/ 45,X.

Azalea = 24 years, high school completed, kitchen assistant, single, height 1 m 58 cm, weight 64 kg, underdeveloped breasts, karyotype 46,XX/ 45,X.

Chrysanthemum = 28 years, high school incomplete, cashier, married, height 1 m 54 cm, weight 65 kg, underdeveloped genitals, lumbar and femoral osteoporosis, karyotype 46X, del Xq(22-qter).

Sunflower = 33 years, high school completed, shop assistant, married, height 1 m 45 cm, weight 42 kg, underdeveloped uterus and ovaries, lumbar and femoral osteopenia, karyotype 45,X.

Primrose = 18 years, primary school completed, student, single, height 1 m 35 cm, weight 31 kg, underdeveloped breasts and genitals, karyotype 45,X/ 46X,r(x).

Lily = 21 years, high school completed, student, single, height 1 m 61 cm, weight 69 kg, underdeveloped breasts, femoral osteopenia, karyotype 46,XX/45,X.

Lotus = 36 years, high school completed, unemployed, single, height 1 m 46 cm, weight 65 kg, underdeveloped genitals, lumbar and femoral osteoporosis, karyotype 45,X.

Tulip = 27 years, university degree in Biomedicine, single, height 1 m 50 cm, weight 40 kg, underdeveloped breasts and clitoris, lumbar and femoral osteoporosis, karyotype 45,X.

Pansy = 26 years, high school completed, cashier, married, height 1 m 44 cm, weight 57 kg, karyotype 45,X.

These women sought out the CAISM due to a situation of primary amenorrhea and, according to their accounts, need to have medical accompaniment all of their lives due to interconnecting factors originating from their TS such as osteoporosis, cardiovascular diseases, renal illnesses among others, as well as their infertility. On the psychological level, the lack of knowledge about the syndrome brings about fears and speculations on other correlations such as mental illness, as well as the psycho-social conflicts such as difficulties in interpersonal relationships; feelings of resignation, rage, impotence, abandon, and states of depression were also unveiled. In order to deal with all of these interconnecting factors they made use of the mechanisms of denial (negation), repression, fantasy, annulment, adaptation and sublimation.

Discussion

Their quotations and significance

We live in a world of symbols; a world of symbols lives in us. Symbol, whose Greek root *sym* signifies together, was for the Greeks a sign of recognition, formed by two halves of a separated object, which, on coming together, is the lost and re-formed unit[14,15]. The visible that points to the invisible[16].

This being the case, the words of the researched women point to important meanings besides the visible fear in the face of the diagnosis of a genetic illness: *"Does TS have any correlation with mental deficiency? On knowing*

about my diagnosis, I went in search of everything about the syndrome since I had believed that mental deficiency was one of the syndrome's characteristics" (Tulip).

These words of anguish illustrate one of the factors that these women associate with TS, which is the fear of, in conjunction with the syndrome, developing a situation of mental deficiency. This patient, even after having sought out information on the syndrome, still looked for confirmation with the researcher in order to acalm down her anxiety coming from an unfavorable and fearful diagnosis and the ignorance of what it really could signify.

Human social groups define normal or stigmatized standards. A person is considered normal when he/she conforms to the previously established standards and the transgression of these standards characterizes stigmatization. The stigma imposes itself on all relationships, like a social construction that is internalized by the majority of people as abnormal. *"By definition it is clear, we believe that someone with a stigma cannot be completely human. Based upon this premise, we make various types of discrimination, by way of which effectively, and very often without thinking, we reduce their chances of life"*[17]. Within the stigmas historically constructed is that of the person with mental deficiency, currently designated "with special needs" (our underlining).

Capitalist society, by way of its rule, privileges "having" in the place of "being". It is a society that does not always value collaboration and solidarity as a counterbalance to unyielding competition and violence. In this type of society, to be a person with special needs is equivalent to not having your due rights recognized, and not even having the equivalent opportunities to those considered normal or non-stigmatized. In view of this, one can infer that the women with TS fear not being fully socially acceptable due to the stigma that their illness brings.

"People picked on me because I am so short..." (Pansy). *"I feel (myself) different because I can't have children like everybody else (in the world)!"* (Chrysanthemum). Feeling different in a massification society that punishes the one who is not equal, stigmatizing them, always brings about an anguished interaction[17]. The body is our most archaic memory. In it nothing is forgotten. Each event lived, particularly in early childhood and also in adult life, leaves a profound scar on the body[16]. So much so that the women researched had demonstrated timid and fearful behavior. *"I would like not to worry so much about what people say"* (Lily). *"I had though that I had some defect"* (Jasmine).

These women fear the consequences of prejudice coming from the diagnosis of a genetic syndrome, to the same extend to which they denounce their own prejudice. Frequently, whatever anomaly brings on a first reaction of repulsion. The anomaly requires, in order to be understood, that one goes beyond the habitual norms of judgment and thus leads to a deeper understanding of the mysteries of the human being and of life.

Responses to falling ill

The responses to the problems created by the illness are socially constituted and directly refer to a world partaking of beliefs and values. The manner in which people position themselves in the face of the illness, conferring upon them significances and developing means to deal with it, are looked upon as experiences of infirmity[18]. Within the researched women these experiences denoted, as well as a lot of pain, feelings of resignation, rage, impotence, low esteem and a framework of depression: *"If it happened this way, it's because it had to be this way..."* (Daisy). *"Why me?"* (Sunflower). *"I have a good job (...) but, I had depression and became very sad, this affected me a lot psychologically"* (Jasmine).

To receive a diagnosis of TS, with all of the clinical interconnections that accompany it, appears to be an unacceptable idea for these women. And their denial is one of their defenses used. *"I had believed that I didn't have TS"* (Jasmine). *"I hid from people what I had, since I didn't want them to know..."* (Violet). To negate painful sensations is a fallback as ancient as the feeling of pain itself. It is very common in young children, since by way of this negation the child realizes wishes that manifest the effectiveness of the principle of pleasure[19].

The repression mechanism is also used as a form of excluding the disturbing idea from the field of consciousness: *"I only remember about it when I come to Unicamp"* (Rose). Not thinking about this avoids having to relive the anguish of being a victim of a genetic syndrome and of feeling different by being infertile. "*I had to know whether or not he would stay with me under these conditions, without being able to have children"* (Orchid). In the end, infertility or the incapacity to procreate can represent a threat to sexuality, in such a manner that bearing children is denied to them and impedes them to feel seductive towards their companions.

"I don't go overboard about this…" (Azalea). In this situation the manifested indifference is no more than a repression and rejection of the unfavorable circumstances with which conscience, or their ego, has to deal. In psychic terms this is equivalent to saying that a diminishing of the ego ideal and consequent increase in narcissism occur, as a manner of resolving the conflict.

"They say that I can't get pregnant, but it's only God who knows this!" (Rose). *"If a have to have (children), if God wants that I have, then I will have!"* (Lotus). Within these words we can infer the use of the annulment mechanism, in so far as they resort to divine omnipotence in order to resolve their problem. Indeed, as this question of religion is highly common, one can contemplate a cultural trait. Nevertheless, if they need to resort to the Supreme and Omnipotent Being, it can be inferred that they feel impotent in the face of the illness and

its consequences. It is the feeling of impotence, in the manner in which it remits back to fragility and human withering away, which brings on frustration and fear. Not to have power signifies being at the mercy of forces that subjugate and that cannot be modified or with which one cannot even fight.

In the same way, fantasy is used as a form of dealing with the situation: *"Once the doctor asked if I had had an accident, but I couldn't remember anything. But a short time ago I remembered that when I was more or less ten years of age I tripped over a glass cupboard, in such a way, with my legs open..."* (Jasmine). It would appear that it is easier to accept that the illness came from an accident in childhood, than to support the idea of an unknown, obscure and mysterious genetic illness. Indeed, those interviewed declared, referring to the diagnosis, which they did not understand "*almost anything of what the doctor had said..."* (Violet, Tulip, Primrose, Daisy).

Was there an error in the professional's explanation or rejection on their part in absorbing the information that had been passed to them? Probably both. Defense mechanisms are psychic processes that look towards reducing whatever manifestation that can place in danger a person's integrity. In this case, the diagnosis of the syndrome, by being excessively unfavorable or threatening, appears to bring about an automatic defensive reaction of the ego looking to maintain homeostatic equilibrium. And, on the other side, one can infer that some professionals have difficulty in clearly explaining to their patient the diagnosis and its consequences, due to their very own defense mechanisms.

The next few quotes point towards the mechanisms of adaptation and sublimation: *"I know that I'll always need to be cared for, I know that taking care of my health is very important and I want to do this"* (Orchid). *"I adopted a baby. It's the child that was born in my heart"* (Daisy). To search for a reasonable means of living with the syndrome and its consequences reveals that the suffering can be elaborated and controlled by these women. The sublimation mechanism is one of the most efficient, since it channels the libidinous impulses towards a socially useful and acceptable posture, as is the case of adoption. In this case, sublimation is considered as the logical and active adaptation of the id's impulses that, in harmony with the superego, satisfy themselves both in benefit of the psychic apparatus and the norms that rule the social context.

We know that the function of the symbol is to give representation to phenomena and, in this manner, to serve as a constructor of becoming human. Life and death, in the same way as loses and gains complement one another in an indissoluble and inherent process. Nevertheless, the experience of significant loses that encapsulate not only situations of physical death but also of symbolic death, can bring about damaging consequences for the person and their family. And this must be approached as the provoker of a crisis, by definition.

Very often the people who are living through the crisis perceive that their resources can be inadequate for this confrontation and they look for external assistance. At other times, they do not perceive their limitations and intensify their suffering or strive to mitigate it at the cost of wearing away their defenses. It is normal for the human being to look to adapt to the difficulties of life. And, in this case, the finality of the defense mechanisms is to reduce or eliminate the anxiety originating from these difficulties. Nevertheless, some mechanisms, in spite functioning as escape valves for anxiety caused by an increase in tension, instead of helping can simple mask the stress and contribute to increasing it.

Therefore, it is important to know the defensive style of women with TS and variants, since it is fundamental for the psycho-diagnosis and for an adequate therapeutic formulation. And in this case, the research's findings serve as a guide in order to consubstantiate the psychological attendance, concurrent with the routine clinical protocol.

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