

Psychological challenges for the cystinosis patient

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There are many psychological obstacles for cystinosis patients, which can be categorised at different levels:

1. Personal and intra-subjective level, that is to say intimate self-questioning, the centrally recurring theme being “why me?” There can be difficulties in “thinking the illness” (and therefore accepting and integrating it), which influences day to day living with this chronic illness.
2. Relational and inter-subjective level, between oneself and others, with a daily self-interrogation as to the views of others, parents and peers. This can generate difficulties in situating oneself-finding one’s place in society, being recognized, and in establishing a “suitable distance” with outside contacts. Relationships with family and friends, the quality of everyday contact with others, and the quality of relationships with doctors, are very important.
3. With regard to the disease, its effect on the body, taking drugs...Compliance problems and cooperation are very important, and sometimes lead to conflict, particularly during adolescence.

Cystinosis patients sometimes experience feelings of anxiety or even despair; feelings of insecurity, disgust, solitude, guilt, and sometimes worthlessness. These feelings are chiefly dependent on affective, corporeal, narcissistic, sexual, social and school-related pre-occupations.

These more or less conscious feelings influence, to a lesser or greater extent, the patient’s life. This leads to multiple reactions relating to the patient’s personality, but also to the particular context and environment. These reactions, whether or not they are overcome, range from excitement to depression, from challenge to defeat, from aggression to inhibition...

All of these feelings can affect the same person in various ways varied way over a period of time. What appears to be important is that cystinosis does not slow down the normal psychological development of the child. Young patients with cystinosis should, above all, preserve their capacity to play and experiment in their surroundings. Cystinosis should be appreciated as being important, which does not necessarily mean that it is serious.

We will look at the different psychological aspects in detail; from the point of view of the patient, depending on age (child, adolescent, adult), and from the point of view of his/her surroundings, taking into consideration family structure and peers.

It should be stated that, from a psychological point of view, there are no specific **mental problems** in children and adolescents with cystinosis. These children and adolescents have a normal personality. In the same way, psychological cognitive function (intelligence, memory, attention, concentration...) and IQ are

identical to that of children without cystinosis. This results in a frequency of psychological problems similar to children without cystinosis.

The main psychological difficulty with cystinosis resides in the difficulty in accepting the disease and living with it, not in suffering from it. It is also the difficulty in being active in a regular and continuous way. The principal issue is to learn to “live with it”, and this takes time. One must accept that it takes time. In trying to achieve this, a key issue is helping and accompanying the child or adolescent, and making sure that he or she does not project or focalize on the disease, for example, “everything is because of cystinosis” or “if that’s the way it is, I won’t take my treatment”. The psychological focus is based on encouragement to mature, followed by maturity, both of which are the keys to accepting cystinosis.

We will define the place and interest of psychological therapy. Of what benefit is psychological support? Whom does it benefit? The child? The parents? Why? When? From the moment of diagnosis? Later? With whom? Psychiatrist? Psychologist? How? - So many questions need to be answered. We will focus on the points which can redress psychological support – identification of genuine psychological problems, resolution of certain situations, stress-alleviation, coping with dead end situations, and helping to understand the dynamics of conflicts.

Finally, we will stress the importance of patient associations, which allow sharing of scientific knowledge, emotions and experiences, as well as permitting a unique and special contact. Psychologically, these associations have a very important role for those living with cystinosis.

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