
Psychological Functioning and Life Experiences in Adults with Congenital Heart Disease

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6.1 Introduction

Although it is relatively unknown in the general population, congenital heart disease (ConHD) is the most common inborn defect. It has an approximate prevalence of 8 newborns for every 1,000 births, and this incidence has remained stable for the last 50 years not varying across countries [1].

Not so long ago, these patients used to carry a very poor prognosis especially when it came to the most severe defects; however, nowadays most of them survive reaching adulthood thanks to the advances and triumphs of cardiovascular medicine and surgery in the twentieth century [2]. For this reason, the number of adults with ConHD is growing rapidly; the growth rate has been estimated to be 5 % per year [3].

Due to this, there is an even more clear necessity of understanding of the impact that the pathology and medical condition has on this population and their psychological functioning during childhood and adolescence [4] and also later on during the adult lives [5] so as to develop more personal and humanized processes, systems, and institutions [6].

In this chapter the literature on psychological functioning adults with ConHD will be presented, later focusing on the life experiences and ways of coping of these patients. Finally a clinical case will be proposed, with the objective of clarifying what the role of the psychologist in the context of congenital heart disease could be.

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E. Callus, E. Quadri (eds.), *Clinical Psychology and Congenital Heart Disease: Lifelong Psychological Aspects and Interventions*,
DOI 10.1007/978-88-470-5699-2_6

6.2 Psychological Functioning of Adults with Congenital Heart Disease

When looking at the research available on the psychological functioning of adults with ConHD, there is often a comparison of this group to a healthy comparison or other populations, and the exploration of the correlation of psychological distress with medical and/or other variables.

Although there are common trends, diverging results have also been obtained, and they can be attributed to the vast differences of instruments utilized, but also perhaps to the different health-care systems and health benefits available in the different countries. In this paragraph we especially refer to a recent review conducted in our group [5] in which the labels of psychopathology, anxiety, depression and hostility, neuroticism, and self-esteem were identified.

The studies by Utens et al. and Van Rijen et al. [7, 8] are examples in which there is a comparison between adults ConHD and the healthy peer group. In the study by Utens and colleagues [7], no differences were found between the mean problem scores of the congenital heart disease adults and reference peers. Only some minor, small, and limited differences were found in two specific problem areas: the “Somatic Complaints” and the “Strange” subscales in the study by Utens.

In the second study of Van Rijen et al. [8], it was outlined that young female patients with congenital heart disease aged from 20 to 27 showed higher levels of psychopathology when compared with the reference group; however, when the mean scores of all separate scales of the patient sample and the reference groups were compared, only small differences on the other informant reports and hardly any differences on the self-reports were found.

A study by Cox [9] also confirmed that adults with ConHD had more favorable scores when compared with the control group; however, it is possible that the control group utilized was inappropriate (a group of patients of the department of orthopedics).

In another study of Utens and colleagues [10], a better functioning of the patients with congenital heart disease was found with respect to the reference group, and it was also confirmed that the population had favorable outcomes on daily and leisure time activities. These results were also confirmed in a study by Van Rijen and colleagues [11] in which a better emotional functioning of the congenital heart disease sample is reported when compared to the reference group.

These results indicate that these patients could have a greater sense of coherence [12] with respect to the healthy population, as also outlined in the chapter on quality of life by Apers, Luyckx, and Moons in this book (see Chap. 7). It is indeed possible that having to face an illness these individuals were made to mobilize more resources as they are growing up when compared to the healthy population and search for solutions or ways of coping appropriate to tackle stress [12].

Another possibility that was suggested was that denial and high achievement motivation could inflate the scores on the self-report data [10, 11]. As we shall see further on in this chapter, appearing to be normal is very important for this population, and this further confirms this possibility.

A study which does not confirm this trend is the one by Brandhagen [13], in which it was specified that adults with ConHD fare worse than the healthy population, showing more psychological distress. It is noteworthy that only 168 out of 463 patients completed the questionnaire.

When it comes to anxiety and depression, in a study by Kovacs [14], the objective was to identify the predictors of anxiety and depression in adults with congenital heart disease. It was indicated that 34 % of the patients resulted as having elevated anxiety. Loneliness and fear of negative evaluation, and not variables relating to the medical conditions of the patient, resulted as being predictors of anxiety.

When it comes to depression, 78 % of the sample resulted as having minimal symptoms, 10 % mild symptoms, and 12 % moderate to severe depressive symptomatology. Loneliness, perceived health status, and fear of negative evaluation resulted as being predictors of depression, and the actual disease severity and functional class did not predict mood severity. Similarly, in a study by Rietveld [15], worse scores on psychosocial adjustment and quality of life were linked to having negative thoughts, irrespective of the medical condition of the patients.

Another interesting thing outlined by the study done by Kovacs [14] is that in the subset of their sample who participated in structured clinical interviews, 29 out of 58 patients met diagnostic criteria for at least one lifetime mood or anxiety disorder. Out of these 39 % had never received any mental health treatment.

In a study by Popelova and colleagues [16] on a small group of patients with persistent cyanosis, depression was associated with older age, worse functional state, and unemployment but independent from the severity of cyanosis. In this case 34 % of the sample resulted as being depressed.

Similarly to Kovacs, in the previous studies of Bromberg and Horner [17, 18], psychiatric interviews were utilized to assess adults with ConHD. In the study done by Horner [18] where 29 patients with complex congenital heart disease were evaluated psychiatrically by both interview and questionnaire, the authors confirm that most of the population were functional in day-to-day life; however, 5 were diagnosed as having panic disorder, and 4 patients met full diagnostic criteria for major depression and had received no psychiatric treatment for these conditions.

Also in the study by Bromberg [17], undertreatment seemed to occur as more than one third of the patients who were assumed to be well adjusted experienced a diagnosable psychiatric disorder. Two patients out of 22 met diagnostic criteria for a general anxiety disorder, and 6 out of 22 patients met diagnostic criteria for a depressive episode.

Most of the literature indicates that there is no relationship between diagnosis, physical fitness or presence of residual symptoms, and worse psychological functioning [7–11, 14, 15, 19], with only few studies suggesting a weak link between physical functional class and psychological functioning [16, 20]. For this reason it is particularly important to explore the possible predictors of psychological distress in this population, in order to incorporate them in clinical practice.

6.3 Factors Associated with Psychological Distress

As specified in the previous paragraph, there are strong indications in the literature that the objective physical condition of these patients is not directly linked to their psychological status. In some studies, other possible variables that could influence psychological distress have been explored [5, 14, 15, 19, 20], and these resulted in:

- Loneliness
- Fear of negative evaluation
- Imposed limits
- Low capacity for physical exercise
- Perceived health status

Perceived health status – In our clinical experience, perceived health status is one of the most important factors which influences how the patients feel about themselves, also because it is an indication that there could be significant gaps in illness knowledge in this population [21]. In a recent study in our center, we specifically wanted to examine the effects of perceived health status by including a numerical rating scale assessing the perceived severity of the patient condition [19]. It was verified that the perception of severity of the patient and not the objective medical criteria, including functional class, resulted as being correlated to psychological distress.

Health perception is the result of the integration of the information and feelings related to the health and of one's limits, the health-care system, the family, and the society. It is influenced by childhood experiences, family relationships, working status, and culture. A lot of information is derived from somatic perceptions (symptoms), from the beliefs and knowledge linked to the disease, and from the information arising from the social environment, in particular the ones given by doctors. All of these components entail both evaluations and information both at a conscious level and an unconscious level, together with the resulting emotional responses, and all these processes influence psychological reactions [22].

Low capacity for physical exercise and imposed limitations and restrictions – As specified in the previous paragraphs, although the general trend in the literature is that the more objective medical conditions are generally not related to psychological functioning, in some studies it was pointed out that physical functionality and the more serious conditions have an influence on how the patients feel about themselves. As we shall describe later, it could be that these restrictions and impositions cause problems in daily life activities, causing problems in the social and work spheres of these patients.

Imposed restrictions resulted as being strong predictors of psychological well-being [20, 23, 24], and these are related to physical activity, lifestyle, and to the possibility of pregnancy. These limits may be imposed by the doctor, on the basis of the functional status and the severity of cardiac disease, and they also may be defined by patients themselves on the basis of subjective experiences, or they may be the result of uncertainties and erroneous beliefs. In fact, from our clinical

experience we have seen that it is possible that patients both greatly underestimate the severity of their condition but also that they feel that the severity of their condition is much greater than a more objective medical evaluation of their situation.

Fear of negative evaluation and social isolation – In a review on the biopsychosocial experiences of adults with congenital heart disease in 2005 [25], Kovacs and colleagues outline how congenital heart disease can impact the social sphere of these patients. It is also important to evaluate the qualitative data available [26, 27] in which there is a further indication of the difficulties of these patients, as we shall describe in more in-depth manner later on in this chapter. From our clinical experience and also the data available, there seems to be a struggle to maintain a “normal” life, and this can be hindered from being evaluated negatively by others and also from having the perception that they cannot participate to social and work activities like everybody else. The importance of these factors is highlighted in the study by Kovacs mentioned previously [14] as both fear of negative evaluation and social isolation are linked with depression and anxiety.

6.4 Life Experiences and Coping Strategies in Adults with Congenital Heart Disease

It is very difficult to generalize about the life experiences of this population, since the different conditions are very diverse. Having said this, there seem to be some patterns of experience and perspective in this broad diversity [28]. Having a complex congenital heart condition can be regarded as having a chronic condition because of the long-term nature of the condition, the uncertainty of its course and prognosis, the signs and symptoms of the condition, and also the restriction on the everyday life of these patients [29].

Qualitative studies which take into consideration the life experiences of this population can give valuable insight on how to further improve psychosocial services for these patients. Indications can also be given on how to improve patient compliance and to strengthen the patients’ coping strategies by taking into consideration the subjective life experiences of these patients.

In two studies conducted on the adolescent population [30, 31], it was outlined how adolescents with ConHD struggle with physical limitations and face social exclusion. The limitations the patients felt were linked to the severity of the condition, and as many as one fourth of adults with ConHD report their parents as being overprotective during their childhoods and adolescence [13, 30, 32]. This could explain why in general these patients tend to live longer with their parents as overprotection could lead to less autonomy [33, 34]. In both researches the theme of being different and the attempts to cope with this emerged strongly.

This central theme of being different from healthy peers also emerges in the qualitative literature pertaining to the life experiences of these patients in their adulthood [26–28, 35–37], and there are often attempts and the struggle to feel normal and to be perceived as being normal by others which can be followed by feelings of ambivalence and also denial of the condition and efforts to exceed their physical boundaries [35].

In one particular older study [37], the problems encountered by the female patients with ConHD were focused upon mainly the concerns regarding fertility, contraception, and pregnancy. Other concerns which the female patients outlined concerned their surgical scars and body size, and they resulted as being uninformed about aspects regarding their condition and reproductive matters. There is an indication that the perception of body image is quite varied in males and in females and that females experience the consequence of scarring and cyanosis more negatively than men, who seem to have difficulties related to their body image during adolescence [26, 27].

Very often the patients refer to their hospitalization experiences, and in a study [38] it was outlined that the patients, their families, and the nurses had different expectations about the management of the physical needs and about the way the patients and the patients' families behaved during acute phases of the patients' illness. These discrepancies were associated with interpersonal conflict, distrust, anxiety, and dissatisfaction with the health care provided.

As outlined in this manual, the involvement of associations relative to this condition is very important. In the chapters by Geier and Campioni Chaps. 12 and 13, the European experience related to this is outlined. In a very interesting article [28], quotations from patients are reported from the Adult Congenital Heart Association in an attempt to address the patients' perspectives in order to outline the implications for care.

When it comes to feedback regarding their state of health, we find two extremities: on the one hand some were told they were "miracle babies" and also at the other end, lost causes, that they would not survive for long, often erroneously. When told that they were lost causes, this often caused an increase in engagement in highly risky behavior and also difficulties to move into adulthood, because of a perception that there was no future to be put at risk or to work to.

In the conditions of moderate and great complexity, it was reported that there was a risk of language-based misperception in understanding if the condition is completely cured or not. There are also often gaps in knowledge regarding what is required to safeguard and optimize their health [21, 39], and this could be related to the fact that there have not been enough efforts to educate patients regarding their condition, the risks associated with it, and the necessity for lifelong follow-up.

Similarly to our experience, it is common for this population to have difficulties when having to go to the "regular" medical system, where the sanitary personnel is not specialized in ConHD. Not only they need to be vigilant because they might be given medication or treatment which could be inappropriate for them, but they report that insisting for specialized care can result in hostility.

ConHD can be considered as an invisible disability, as quite often it is not evident to others that the person has this condition. The adults often report that friends and family do not really understand their restrictions and that sometimes they also believe that changes in various responsibilities which could be cumbersome are not really required.

Other difficult situations the patients have to face are making difficult decisions regarding their medical care, referral for surgery, and in those occasions when they

are informed that there is nothing left to be done to improve their health [18]. End of life care in this population is a particularly sensitive topic which will be developed in detail by Kovacs and colleagues in this book (Chap. 8).

The patients have also reported that there were occasions in which information is withheld from them and they got to know about their real condition at the onset of new problems. This made it much harder for them to adapt, also because of feelings of betrayal and anger.

Finally, as already specified before, having this condition does not only have negative consequences but there are also some benefits. These include receiving special attention, increased resilience and maturity, and also gaining a clearer sense about the meaning of life. The ongoing awareness of one's mortality can lead to an increased appreciation of life, more clarity of purpose, and better decision making [40].

6.5 Coping Strategies in Adults with Congenital Heart Diseases

From the literature already described, it can be seen that although this population faces many challenges, there is a good social adaptation. In the qualitative study of life experiences already mentioned by Claessens [27], the personality of the subjects is related to the use of particular strategies: in particular, an internal locus of control is related to more active strategies, while adults with an external locus of control seem to be more passive, with a consequent lower adaptation and resignation to feel different, with the consequent risk of social isolation.

In this study, the "process of normalization" is also outlined, understood as dynamic process and management strategy through which the sick person and the family strive to live in a manner similar to that of the healthy peers. This process has its origin from the experience of feeling different from healthy subjects, and it is influenced by the age of the subject and the following factors:

- The environment (ConHD understood as disabilities or realistic vision of the disease)
- The health-care system (medical treatments, approach to the care, and information)
- The personality of the subject

In particular, a realistic view of the disease and the limitations imposed by it, with adequate information, seem to favor the adaptation and coping to the limits imposed by the condition and a better integration of illness management in everyday life.

In a similar study by our group, Callus et al. [26], we also consider the different types of coping strategies utilized to deal with their condition, applying to them the conceptual framework of Moos and Holohan [41]. The patients themselves describe avoiding coping strategies they had utilized previously as not being adaptive,

especially when it comes to the defiance of limits. Other avoidance strategies, such as accepting the inevitable and the utilization of humor, did seem to be adaptive from what the patients described. Denial on the other hand might have been functional at times, such as before an intervention, but it seemed to have consequences anyway, and often it leads the patients not to acknowledge cardiac-related symptoms which could have put their health in danger.

It is interesting to note that the patients who underestimated their conditions the most are the ones who admit ignoring having the condition for many years avoiding medical checkups or defying the therapeutic indications completely. This was also due to the family background they were brought up in and their lack of education and cognitive resources. When dealing with their condition, they describe utilizing mostly cognitive and behavioral avoidance coping strategies.

According to the study of Van Rijen and colleagues [42], the styles of coping of this population resulted as being very similar to their reference groups, with very few differences. The female patients reported lower active problem solving, and the males showed more favorable styles of coping compared to their peers, such as higher seeking of social support, lower passive reaction patterns, and lower expression of negative emotions. Although the adults in this study reported receiving less social support, they also highlighted a lower discrepancy between desired and received social support, indicating feelings of independence in these adults.

In other studies possible defense mechanisms were mentioned [10, 35, 43], in particular the denial of the problems associated with the cardiac malformation and compensation. In particular denial can be manifested in different ways, from the extreme denial of having a condition to the enactment of various strategies to relieve stress. Horner [18] highlights how denial can be adaptive, but that it can also not be functional, worsening the condition of isolation and anxiety.

Strategies for coping in patients with ConHD should be more explored as they are critical in predicting the adaptation to the disease. The education should be customized and designed to replace the responses of passive coping with a more active style of coping, which increases the perception of control over heart condition on the part of the patient and to promote better integration of the disease in the context of life.

In the next paragraph we report a clinical case study of psychological support being provided postoperatively, in an attempt to make a connection between the issues mentioned in the literature and the subjective life experience of one patient.

6.6 The Role of the Psychologist: A Clinical Case Study

Gloria is a 44-year-old woman hospitalized at the IRCCS Policlinico di San Donato Milanese University Hospital for an aortic valve replacement. She is diagnosed with a congenital aortic valvular insufficiency, ascending aorta aneurysm, and pulmonary atresia with an interventricular septum defect.

At the age of 12, she underwent her first cardiac surgery in Birmingham with a positive outcome. Before this surgery, a mild aortic valvular insufficiency was discovered which was followed with regular outpatient visits. At the various controls,

a progressive deterioration of the valve insufficiency and aortic aneurysm dilatation of the ascending aorta associated were discovered, and the patient reported greater fatigue.

At the time of hospitalization, Gloria was referred to the psychological team by the medical staff because she was in a lot of pain due to the fact that she had to keep on the drains for a longer period after the cardiac surgery. The role of the psychologist in psychocardiology is tackled by Prof. Compare and colleagues in this book (Chap. 9), and for these reason here I just give a summary of the role of the psychologist when it comes to this population. Psychologists could be a useful figure in accompanying adults with ConHD patients in the various phases of the illness:

- Coping with becoming aware of the condition or changes in cardiac status [18, 44, 45]
- Accompanying the patient in the difficult pediatric-adult transitions [45, 46]
- Adjustment to cardiac devices and surgical preparation [47–52]
- Maximization of adherence and behavioral modification [53, 54]

It is possible that referrals to psychologists can be initiated by both the patient and the medical staff. It is always important that a psychological (or even a psychiatric) referral is made with the patient's knowledge and to enquire with the patient initially how they feel about receiving this kind of support if it is to be suggested. It is essential to clarify appropriate referral indications with the psychologists working with an adults with ConHD team [45]. Referral to other specialists such as psychiatrists [55] and experts in substance abuse (especially since it has been linked to unsuccessful transitioning from pediatric to adult care [46]) could be necessary in particular cases.

In Gloria's case, she welcomed the possibility to receive psychological support. During the first session, when she is asked how she is feeling, she says that she fears that her reaction to pain and discomfort is not appropriate. When exploring what these feelings are linked to, she explains how everyone she knows, including her husband, encourages her to be strong and to be positive and not to complain and that even if she would like to do so, she is not managing, and for this reason she feels very discouraged.

Firstly, her experience is validated and she is encouraged to talk about how she feels, without passing judgment about this. Gloria welcomes the possibility to be able to express herself, and she also talks about how she felt in some previous hospitalizations.

It is explained to her that it may happen that our loved ones struggle to support our pain, and they try to encourage us. By doing so, they unwillingly do not truly accept and listen to our suffering, and this causes frustration in a person who is in difficulty, who does not feel understood.

Gloria's husband is concerned about her well-being, and he asks the staff about how she is doing, also when it comes to her psychological well-being. It is specified to him that it is quite normal to be experiencing difficulty in this kind of situation, that there are no right or wrong ways to feel and that it is particularly important for cardiac patients who have undergone surgery to feel that there is support without

judgment on how they should be feeling. After this, some indications are given to him on how to be supportive, specifying that it is common to use encouragement in an attempt to help others, but that this can be counterproductive and that it is important in this moment just to listen and to be there for his wife.

In the following sessions with Gloria, the importance of legitimizing one's feelings and giving them relevance is explored – and it is specified that it is an important first step if we also want others to give importance and relevance to what we are feeling and going through. Once she is able to express how she feels regarding her pain and her frustration, Gloria asserts that she feels much better.

The next thing she talks about is her surgical scar. As specified previously, this is an important theme especially for women. She asks feedback about the appearance of her surgical scar and expresses difficulty when it comes to looking at herself. As specified previously, the theme of the surgical scar is important, especially when it comes to females. It is suggested to Gloria to look at herself in the mirror and to express how she feels, welcoming it, while being supported.

She accepts to do this and when she looks at herself, she is very moved. She starts crying and describing on how she has always felt inadequate and that she struggled to accept her illness when she was a little girl. It is suggested to her that the difficulty to look at herself is also linked to these experiences and of seeing herself as being ill; the scar could be a symbol of all of this. Gloria confirms that this is so, and it is agreed that what it means to be ill for her and her feelings of inadequacy are to be explored in the next sessions.

Gloria describes her difficulties at school when she was a child, especially in middle school, the year before she had her first cardiac surgery. She talks about her feelings of inadequacy which were linked to the physical limitations and to the consequent sensations of shame when she compared herself to others in her class. She was particularly ashamed of the fact that she had to be accompanied by her mother in outings because her teachers would not want to take responsibility for her because of her condition. In this case study, it is clearly confirmed that the physical restrictions and the impositions from others have an important impact on the psychological well-being of these patients.

Referring to this in particular, she talks about an episode in which a physical education teacher did not want to include her in the activities because she had this condition and not only – he asks her in front of everyone if she is ill in an indelicate manner. Gloria remembers how she would have liked to become invisible in that moment. She tells everyone to stop looking at her, making the situation worse. Since then she says she will always try to make herself invisible, so as not to be humiliated in front of others.

Remembering about her childhood, Gloria also talks about another physical education teacher, who used to allow her to do physical activity, whatever she managed to do, telling her not to insist if she felt tired. A comparison is made between the two situations, and an agreement is reached about the fact that it is not the condition which made her feel inadequate, but the attitude of some adults who were responsible to take care of her when she was a child which was not appropriate.

After that, it is suggested to Gloria to describe the traumatic event in the present, and Gloria relives the experience and she is very touched emotionally. After that, she is asked to relive the experience as an adult and she said that she would answer back to the teacher. The session is concluded by differentiating these two parts, with the awareness that there is a part of her which feels vulnerable, but also another part which is more adult and which has the capacity to defend itself.

Finally, the possibility that this feeling of inadequacy and shame could have been linked to these memories because they had never been shared with anyone is explored. In the final two sessions, the themes which emerged are recalled to increase Gloria's awareness on her personal growth. She is very happy to have had the possibility to explore how she feels and her life experiences. She affirms that she probably would not have undertaken psychological treatment, had it not been offered to her in this context. This also confirms the possibility of the undertreatment of psychological distress in this population, as mentioned earlier, which is present in the general population but which could be especially relevant in adults with congenital heart disease, where there is a strong need to feel and seem "normal," often having to face many adversities.

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