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**CLINICAL PSYCHOLOGY AND ADULTS WITH
CONGENITAL HEART DISEASE**
DISEASE PERCEPTION, HEALTH RELATED QUALITY OF LIFE
AND LIFE EXPERIENCES

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Chapter 1

General Introduction

1.1 Humanisation in health care contexts

In the last decade there has been a greater attention to the experience of patients receiving healthcare (Todres, Galvin, & Dahlberg, 2007). The current research is developed in this context; the sanitary personnel in the Policlinico San Donato Hospital has become aware that these congenital heart disease (CHD) patients and their relatives necessitate a lot of support because of many factors; uncertainty, frequent hospitalisations, restrictions and difficulties in work and social spheres (Kovacs, Sears, & Saidi, 2005; Verstappen, Pearson, & Kovacs, 2006).

In this period, there has also been a renewed interest in the concept of “citizenship” which is opposed to the one of “being a consumer” (Ridley & Jones, 2002). The phrase which defines this concept is “we are all in this together” and also the fact that we have responsibilities as well as rights (Turner, 1990). Therefore, placing a greater emphasis on citizenship healthcare would mean that our health and well-being belonged to us, and that the systems and consultation process would be designed around this. This is so because although technological progress has brought many benefits, it has also entailed the risk of potentially developing less personal and less humanised processes, systems and institutions (Todres et al., 2007).

The potentially dehumanising implications of technological progress have been historically discussed by many authors (Habermas, 1990; Heidegger, 1966; Marx, Moore, Aveling, & Engels, 1954). In particular, Foucault & Sheridan (Foucault & Sheridan, 1976) analyzed how the process of advancement in technology could result in depersonalizing and dehumanizing forces, especially when it comes to medical institutions. Already in those years he argued convincingly that the medical and technical conceptions of health and illness became a powerful language used in a way which perpetuates the depersonalizing and dehumanising practices of care.

In an increasing number of qualitative research studies, the need for a more humanised approach to treatment is emerging (Galvin, Todres, & Richardson, 2005; Todres, Fulbrook, & Albarran, 2000) and this need also emerges in the literature on Adults with Congenital Heart Disease (ACHD) and also in the empirical studies outlined in this dissertation.

1.2 Incidence and aetiology of CHD

CHD is the most common inborn defect, with an approximate prevalence of 8 newborns for every 1000 births; this incidence has remained stable for the last 50 years and it does not vary across countries (Hoffman & Kaplan, 2002). Previously these patients used to carry a very poor prognosis, however nowadays most of them survive reaching adulthood. In fact, the number of ACHD is growing rapidly due to the advances and triumphs of cardiovascular medicine and surgery in the 20th century (Gatzoulis, Swan, Therrien, & Pantely, 2005); the growth rate has been estimated to be 5 percent per year (Brickner, Hillis, & Lange, 2000a). Especially for this reason it is important to focus research efforts to improve the handling in these patients, since they are a population which is on the constant increase.

Although CHD should by definition be present since birth, some conditions are not usually detected in early life (such as moderate atrial septal defect) and others are only anatomically present in later years with a latent predisposition prior to this; even though they are not strictly congenital they are included in this group. In most cases CHD is diagnosed before the patient reaches one year (60%), most of the remaining patients are diagnosed in childhood (30%), and the remaining receive their diagnosis when they are over 16 years of age (10%) (Gatzoulis et al., 2005).

Both genetic and non-genetic factors are responsible for the aetiology of CHD, however genetic factors play a much greater contribution. Out of the most common genetic conditions which are associated with CHD are Trisomy 21 (Down syndrome), DiGeorge Syndrome (CATCH 22) and Williams syndrome (Gatzoulis et al., 2005).

These patients pose a serious challenge because CHD include a large variety of diverse anatomic malformations of varying severities which differ according to their natural history and also the degree of anatomic repair received. Because of this, they should ideally be treated by cardiologists trained in both paediatric and adult

cardiology, who also receive training in the special problems presented by this population (Gatzoulis et al., 2005).

Out of these patients, one third is considered to have a simple condition, however the majority have lesions of great complexity. Most of them will also have lifelong ongoing CHD and other medical needs (Gatzoulis et al., 2005). In fact, more than a third of the patients is born with “critical heart disease” involving life threatening malformations which necessitate palliative or corrective surgery in early life (Marino, Bird, & Wernovsky, 2001; Samanek, 2000).

CHDs can be classified according to cyanosis (Brickner et al., 2000a; Brickner, Hillis, & Lange, 2000b) severity (Kovacs et al., 2005; Warnes et al., 2001) or underlying structural characteristics (Gatzoulis et al., 2005). When it comes to severity, the defects have been categorised in three classes; simple lesions, moderate lesions and severe lesions; their prevalence being 51%, 26% and 23 % respectively (Hoffman, Kaplan, & Liberthson, 2004).

1.3 Research context and rationale

Kovacs et al (Kovacs, Silversides, Saidi, & Sears, 2006) have specified that there are three factors which affect the possible involvement of a psychologist in an ACHD team; these factors shall be utilised in order to give a description of the research context and rationale of the current research.

1.3.1 The commitment to the psychosocial concerns of this population;

The-San Donato Policlinico Hospital is the leading heart surgery centre in Italy and one of the world leading centres in the research and care of children heart patients. In 2006 it was recognised as a Scientific Institute for research, hospitalisation and health care (IRCCS) for cardiac and vessel illnesses, confirming a vocation in didactics and research which has carried on for twenty years.

Amongst its research lines, the first one regards “Heart and vascular congenital pathologies” and amongst the objectives there is the study of CHD with particular reference to the psychosocial aspects of the patients treated for this condition. In fact, since June 2006 the hospital has invested in the hiring of 2 psychologists, working on the psychosocial aspects of these patients. This is a good step forward because as

stated by Kovacs et al. (2006) it is often hard to find an adequate reimbursement for psychology, in both the American, Canadian and European contexts.

1.3.2 The size of the ACHD program;

The San Donato centre is one of the few centres in Italy which is structured with the presence of a centre which studies CHDs in adults, where a multidisciplinary team made up of the paediatric cardiologist, the cardiac surgeon, the psychologist, the internist, the haematologist and sometimes the gynaecologist work together to deal with the clinical problems caused by this condition.

Approximately 400-500 interventions in the field of paediatric cardiology and 600 cardiac catheterization procedures a year are performed, making this hospital setting an adequate one to start this research program (I.R.C.C.S. Policlinico San Donato, 2008).

1.3.3 The availability of psychologists who have specialised cardiac experience;

My colleague Dr. Emilia Quadri and I have been researching and interacting with this population since June 2006, in order to gain the necessary expertise to effectively handle these patients, and to give our contribution to the multidisciplinary team operating in this hospital. This doctoral dissertation is also an effort in this direction.

1.4 Dissertation outline and objectives

This dissertation has three main goals: (1) to verify if ACHD patients rate the severity of their condition accurately (2) to investigate the links between disease severity as rated by the patients and the physician, health related quality of life (HRQoL) and illness behaviour (including coping) and (3) to provide indications in the psychological handling of ACHD patients.

In **Chapter 2**, the role of the psychologist in the care of ACHD will be outlined, starting from the definition of Clinical and Health psychology, and the description of the areas which contributed to “Psycho-Cardiology”. Some differences between acquired and congenital heart disease will also be outlined.

In **Chapter 3** the literature regarding the psychosocial aspects of ACHD patients will be presented, paying particular attention to research on Quality of Life (QoL) and psychological functioning. The social issues which this population face will also be addressed.

The quantitative research regarding the relationship between disease severity perception and evaluation, and HRQoL and illness behaviour will be presented in **Chapter 4**, linking it with the research presented in the previous chapter.

In **Chapter 5**, there will be a focus on the literature which takes into account the ACHD patients' life experiences and perspectives, beginning with coping with chronic illnesses and disability in general to focus on the difficulties and also the gains, such as increased sensibility and maturity, which result from having CHD.

The qualitative research regarding the life experiences and coping strategies of ACHD patients will be presented in **Chapter 6**, once again linking what emerges from the unstructured interviews with the literature previously outlined in both chapters 3 and 5.

Finally in **Chapter 7**, the final discussions and conclusions will be presented; the main areas which constitute "Psycho-Cardiology" will be utilised as a framework to present implications for the psychological handling of ACHD patients which emerged from both the literature taken into consideration and the empirical studies presented.

In summary, the following questions have served as guidelines when carrying out this dissertation:

1. Do ACHD adequately rate the severity of their condition?
2. What are the consequences of disease severity over and underestimation?
3. What is HRQoL related to during hospitalisation?
4. How do ACHD patients perceive themselves in their illness?
5. What are the life experiences of these patients and how do they cope with their condition?
6. And finally, how can the clinical psychologist give a valid contribution in the handling of these patients in a clinical setting?

Chapter 2

Clinical Psychology, “Psycho-Cardiology” and CHD

2.1 Clinical Psychology and “Psycho-Cardiology”

Clinical psychology includes the scientific study and application of psychology for the purpose of understanding, preventing, and relieving psychologically-based distress or dysfunction and to promote subjective well-being and personal development (American Psychological Association, 2008; Plante, 2005). Psychological assessment and psychotherapy are central to its practice, although clinical psychologists also engage in many other activities, such as research, teaching, consultation and forensic testimony (Brain, 2002).

Amongst its areas of specialization, one can find health psychology, psychosomatics and hospital psychology, where the psychologist contributes in interventions aimed to maintain and promote health, identify the etiological and diagnostic correlates and to analyze and improve health care and to enhance public health (Imbasciati, 2000).

Interestingly, one of the fastest growing area seems to be health psychology, which is reflected by the fact that hospitals have been the fastest-growing employment setting for clinical psychologists in America in the past decade (Benjamin, 2005), and the development of “Psycho-Cardiology” occurred in this field.

It is important not to restrict the care process only to the organic dysfunction but also to anything that is or could be related to disease itself (Molinari, Bellardita, & Compare, 2006). This stance is particularly clear in the Biopsychosocial (BPS) Model presented by George Engle as a holistic and alternative to the prevailing biomedical model which had dominated industrialized societies since the 20th century (Engel, 1977).

Whilst acknowledging the importance of the mainstream of biomedical research in the advances of medicine, he criticized its excessively narrow focus for the leading clinicians to regard patients as objects, excluding the possibility of studying

subjective experience scientifically. In fact the proposal of the BPS model was not only a scientific one, but also a fundamental ideology that tried to reverse the dehumanization of medicine (Borrell-Carrio, Suchman, & Epstein, 2004).

“Psycho-Cardiology” is not intended to be a new discipline on its own but rather, it is an effort to see how psychology can contribute in the prevention, treatment and rehabilitation of patients with cardiac disease (Molinari et al., 2006). In this perspective, psychological and cardiac issues are not seen separately and there is a belief that a multidisciplinary approach is necessary for the wellbeing of the cardiac patient. It is also necessary that the knowledge and skills derived from training in clinical psychology to be tailored to the needs of cardiac patients, and also to cardiac patients with different diseases as different cardiac conditions also have different implications.

2.2 The historical development of “Psycho-Cardiology”

Historically, the first studies that promoted the growth of “Psycho-Cardiology” investigated the link between personality factors and cardiac illness. Friedman and Rosenman identified a complex sequence of behaviours and affects which they categorised into the category “type A behaviour pattern” (TABP) in the fifties (Friedman & Rosenman, 1959, 1974).

Although a unanimous conclusion about the relationship between TABP and cardiac disease was not reached, their studies were an important turning point because they demonstrated that it was possible that specific personality traits may influence cardiac health negatively. More recently a new personality pattern was associated to cardiac disease; the Type D personality or “Distressed Personality” (Denollet, 1991, 1994, 1997, 2000). Although Type A and D personality have different characteristics it could be that social desirability is a common underlying feature (Molinari et al., 2006).

In the sixties other types of research focused on behavioural risk factors particularly studies focused on health-related lifestyle and addictive behaviours, which lead to cognitive-behavioural programs aimed to modify risk factors (Ezzati & Lopez, 2003; Fitzgerald, Prochask, & Pransky, 2000; Scherwitz & Ornish, 1994; Sebrechts, Falger, & Bar, 2000).

Finally another research current which emerged was the association between cardiac disease and psychopathology, especially focusing on anxiety, depression, excessive workload, stress and social isolation (Molinari et al., 2006). Psychological distress is commonly reported after patients receive a cardiac diagnosis, and a substantial amount of them (20-50%) report having adjustment difficulties (Carney, Freedland, Sheline, & Weiss, 1997). The relationship between psychosocial factors and the occurrence and course of cardiac disease is considered to be relevant and plausible (Rozanski, Blumenthal, Davidson, Saab, & Kubzansky, 2005) and these factors have also been investigated in the CHD population, focusing on the course of the illness since the occurrence is genetic (Kovacs et al., 2005).

2.3 Basic elements in “Psycho-Cardiology”

Currently there is no specific training program in the field of “Psycho-Cardiology”, however some theoretical and methodological approaches typical of clinical psychology could form part of a good theoretical framework for this field (Molinari et al., 2006):

1. Carl Rogers’ concept idea of “**patient focused therapy**” (Raskin & Rogers, 1989). He believed that the client only needed three things from a clinician to experience therapeutic improvement—congruence, unconditional positive regard, and empathetic understanding (McMillan, 2004). This theoretical framework emphasizes the fact that the clients have the necessary resources to reach their full potential and that therefore the psychologist should assume a non-directive stance. Surely these qualities can help the cardiac patient to open up and increase therapy compliance, however sometimes a more directive approach is necessary due to particular patients and contextual and temporal limitations.
2. Contributions of **Positive psychology**, the scientific study of human happiness and well-being. Its focus is the increase of the positive experience of life through optimism and the development of personal traits like courage, perseverance and altruism (Linley, Joseph, Harrington, & Wood, 2006; Snyder & Lopez, 2001). In this field there is an emphasis on the role of individual resources and abilities, instead of focusing on what is wrong and dysfunctional; in fact this field has also encouraged an increasing interest in

research on coping (Livneh & Martz, 2007). Its contributions in “Psycho-Cardiology” are the promotion of QoL for patients and their family members.

3. Cognitive-behavioural therapy (Emmelkamp & Oppen, 1998; Hollon & Beck, 1994). The main goals of many cognitive behavioural therapies is to find out and identify the biased, dysfunctional ways of relating and reacting of the clients and to help them transcend these in ways that will lead to increased well-being through different methodologies (Beck, Davis, & Freeman, 2007). The main elements of the therapeutic process in this model are:

- a. The definition of how and when dysfunctional behaviour is enacted;
- b. Identifying any ineffective strategies utilized to face the problematic situation;
- c. Discovering and utilizing problem or emotion oriented effective strategies (coping strategies) (Meichenbaum, 1997).

This therapeutic model/framework is particularly useful when treating cardiac patients because usually the intervention is brief and focused and both patient and therapist play an active role in setting goals and in identifying specific cognitive patterns that may generate problems. For this reason it could be useful to try to reduce risk factors and unhealthy behaviours, although the working alliance must always be taken into consideration in order not to provoke resistance in these patients. This might occur especially if the psychologist adopts a “psycho-educational” stance, focusing on the problems and not taking into consideration the patients’ perspectives (Molinari et al., 2006).

4. Systemic family therapy. In this model the central focus tends to be on interpersonal dynamics, especially in terms of how change in one person will affect the entire system (Bitter & Corey, 2001) The occurrence of any destabilizing event, including the diagnosis of a cardiac diagnosis or any other serious illness causes a break down in the existing family system. This model is useful when the family system does not manage to re-establish a “homeostasis” after a cardiac event, with the consequence of increasing emotional distress and conflict resulting directly or indirectly in a lower quality of life for the patient’s family and a stress on the patient’s medical and psychological condition (Molinari et al., 2006).

2.4 The role of the psychologist in cardiac illnesses: from acquired to congenital heart diseases

Within an approach in which there is a focus not only on the pathological processes but the person as a whole, the psychologist can give a precious contribution to a multidisciplinary team. In particular, they can help the cardiologist in the following ways (Allan & Scheidt, 1996):

- By offering support to patients during and after hospitalisation when it comes to therapeutic alliance, modifications in lifestyle and by reprocessing the trauma caused by being a “victim” of the cardiac event;
- The personalization of therapy, which facilitates adherence and diminishes hospitalisations due to recurring illness;
- The improvement of the communication between doctor and patient;
- Carrying out differential diagnosis when necessary:

Cardiac rehabilitation has been defined as a multi-component and coordinated intervention aimed to optimise the cardiac patient’s physical, psychological and social functioning and to reduce morbidity and mortality (Leon et al., 2005). The Italian guidelines for psychological activity in rehabilitative cardiology identify the different moments which characterise the interactions between cardiac patients and psychologist, summarised in the following 5 phases (Sommaruga et al., 2003):

1. **Selection:** the patients have the right to ask and obtain a psychological consultation; if the organisational resources allow for it, the patients who could require a psychological intervention should be identified, and the patients’ requests should also be guided towards the most appropriate care process;
2. **Entry:** this defines the first contact between the patient and the psychologist;
3. **Assessment:** the objectives are to identify the possible psychological problems, the needs and the resources and to gather all the data necessary to identify the appropriate way of intervening. In this phase the focus of the attention should be getting to know the behaviours and habits which are not functional to the health of the patient as opposed to psychopathological aspects;
4. **Intervention:** the objective of the psychological intervention in rehabilitation cardiology is to help the patients and their families to:

- Recognise and express their own emotions regarding the illness;
- Identify and carry out strategies in order to control risk factors and to modify their lifestyle;
- To implement the correct self management of rehabilitation treatment based on the characteristics of the individual;
- To re-obtain a satisfactory quality of life.

The following effective psychological interventions have been outlined in the literature (Bennett, Cordes, Westmoreland, Castro, & Donnelly, 2000; Dusseldorp, van Elderen, Maes, Meulman, & Kraaij, 1999):

- Group or individual educational interventions;
- Individual or group counselling;
- Stress management interventions;
- Individual and group psychotherapy (mainly cognitive-behavioural);
- Self-help interventions.

5. **Follow-up:** in this phase the patients' satisfaction regarding their needs is evaluated and there is an assessment of the presence of the conditions which may necessitate continuity of care.

CHD varies from acquired cardiac illness in that its onset is genetic, and therefore this population is much younger, and as stated before the adult population is always on the increase; in fact there are more adults than children living with CHD (Webb, 2005). Several authors have already emphasised the importance of recognising and managing the potential psychosocial consequences of growing up with CHD (Kovacs et al., 2005; Moons, De Geest, & Budts, 2002) and international working groups have also outlined the necessity of including specialised mental health care for ACHD patients (Connelly et al., 1998; Deanfield et al., 2003; "Summary of recommendations--care of the adult with congenital heart disease," 2001) ("Care of the Adult with Congenital Heart Disease. Presented at the 32nd Bethesda Conference. Bethesda, Maryland, October 2–3, 2000," 2001).

Despite these recommendations it seems that the under-diagnosis and under-treatment of psychosocial concern are present in ACHD patients (Bromberg, Beasley, D'Angelo, Landzberg, & DeMaso, 2003); for this reason it is important to include a psychologist in ACHD teams. Kovacs and colleagues (Kovacs et al., 2006) identified three main domains in which clinical health psychologists can contribute in an ACHD

team: provision of clinical services, multidisciplinary research and professional education.

2.4.1 Provision of clinical services

As specified in the next chapter, ACHD can affect psychological functioning and it is possible that the patients are more sensitive to heart-focused anxiety and bodily sensations due to their condition (Eifert, Zvolensky, & Lejuez, 2000; Rietveld, Karsdorp, & Mulder, 2004; Utens, Bieman et al., 1998). Psychologists can help in the reduction of excessive self monitoring and reduction of cardiac anxiety through psycho-education and behavioural strategies. They can also help patients to deal with family and peer concerns through social skills training and guidance in communication strategies (Kovacs et al., 2006)

A list of six clinical strategies in order to maximise psychosocial care in ACHD patients has been previously identified in the literature (Kovacs et al., 2005):

1. **Demonstrate increased psychosocial awareness:** in settings which work with ACHD patients
2. **Initiate proactive discussions:** trying to avoid important topics will harm the patient in the long run, and initiating discussions allows them to feel they can ask questions and that what they are going through is common.
3. **Screening;** Employing the 4 A's to detect and manage psychosocial issues:
 - a. Ask the patient about specific challenges,
 - b. Advise the patient on common challenges and how to manage them,
 - c. Assist the patient through psychological interventions such as support and brief problem solving, and
 - d. Arrange referral when the patient is evaluated as needing a mental health specialist.
4. **Build improved clinical strategies**
5. **Consider regionalization of clinics**
6. **Increase longitudinal research:** especially when it comes to psychosocial interventions which as specified before, are missing in this field.

Psychologists could be a useful figure in accompanying ACHD patients in the various phases of the illness;

- **Coping with becoming aware of the condition or changes in cardiac-status:** they can be useful to help alleviate psychological distress which can arise for a variety of reasons; one of which is coping with one's medical condition, especially since many of ACHD patients require lifelong medical follow-up (Warnes, 2005) even though they consider themselves as being cured. Patients can also feel angry if they suddenly get worse after many years of relative stability (Horner, Liberthson, & Jellinek, 2000). Some patients also learn about their condition in adulthood and could have a hard time adjusting to this (Kovacs et al., 2006);
- **Accompanying the patient in the difficult paediatric-adult transitions:** the transition from paediatric to adult care results to be very difficult for many ACHD patients, with many adolescents who do not manage to successfully transition to adult care (Reid et al., 2004). Psychologists can provide support and individual consultation to those patients experiencing a difficult transition through strategies such as empowerment (Kovacs et al., 2006);
- **Adjustment to cardiac devices and surgical preparation:** It is possible that some patients have difficulty with the implantation of cardiac devices such as the ICD, especially if they are young (S. F. Sears, Jr., Todaro, Lewis, Sotile, & Conti, 1999), and cognitive behavioural strategies have been shown to be useful to enhance the confidence and QoL of ICD recipients (S. F. Sears, Kovacs, Azzarello, Larsen, & Conti, 2004). Psychological factors can have a negative impact on surgical recovery (Kiecolt-Glaser, Page, Marucha, MacCallum, & Glaser, 1998) and some meta-analytical studies have demonstrated that psychological and psycho-educational preparation for surgery can have a positive impact on surgery outcomes (Devine, 1992; Johnston & Vogege, 1993). Psychologists can provide preparation techniques which improve the reduction of anxiety in cardiac patients which have to undergo a surgical procedure (Seskevich, Crater, Lane, & Krucof, 2004);
- **Maximisation of adherence and behavioural modification:** it is especially important for cardiac disease patients to take care of their diet, to exercise appropriately and not to indulge in risky health behaviours which can compromise their health. Psycho-education seems to be helpful with it comes to lifestyle

behaviours in patients with coronary heart disease (Dusseldorp et al., 1999) although there are no specific studies regarding this in ACHD patients. These patients seem to engage less in physical activity (Reybrouck & Mertens, 2005) and behavioural strategies could be utilised to adopt and maintain physician approved physical activity regimens and also to adhere to the physician's recommendations;

- **Dealing with anticipatory grief and mortality:** It is also possible that the patients of this population have an increased awareness of mortality and a psychologist could help be providing support, dealing with anticipatory grief and working on strategies to communicate effectively with relatives and sanitary personnel.

Referrals to psychologists can be initiated by the patient or the physician. Sometimes ACHD patients enquire themselves if a psychologist available, at other times the physicians broach the topic with their patients. It is always important that a psychological (or even a psychiatric) referral is made with the patient's knowledge. It is essential to clarify appropriate referral indications with the psychologists working with an ACHD team (Kovacs et al., 2006). Referral to other specialists such as psychiatrists (Bassett et al., 2005) and experts in substance abuse (especially since has been linked to unsuccessful transitioning from paediatric to adult care (Reid et al., 2004)) could be necessary in particular cases.

2.4.2 Multidisciplinary research

Psychologists can contribute in the conduction of multidisciplinary research in ACHD patients, especially in the following areas (Kovacs et al., 2006):

- **Development of ACHD-specific measures:** there have been many studies using different instruments in this population, and there is a need for the creation of specific psychosocial measures developed for this population ("Care of the Adult with Congenital Heart Disease. Presented at the 32nd Bethesda Conference. Bethesda, Maryland, October 2–3, 2000 ", ; Deanfield et al., 2003). There already have been some attempts in this direction (Kamphuis et al., 2004), although additional measures are required in order to address the unique concerns of this population, as the psychosocial measures developed for acquired cardiac disease do not capture the full ACHD patient experience (Kovacs et al., 2006).

- **Longitudinal psychosocial assessment:** there are few of these studies present in the literature and more are required to understand how the population deals with the various issues through time.
- **Evaluation of psychosocial interventions:** no trials yet exist when it comes to psychological interventions for ACHD patients (Lip, Lane, Millane, & Tayebjee, 2003) and since there are few specialised centres it is also feasible to think about how to provide long distance support and how to evaluate it ("Care of the Adult with Congenital Heart Disease. Presented at the 32nd BethesdaConference.Bethesda, Maryland, October 2–3, 2000 ").
- **Evaluation of medical interventions and the relationship between mental and physical health:** psychologists can provide insights on the effectiveness of medical interventions by exploring how the patients feel before and after, especially in the domains of quality of life and psychological functioning. It is also important to assess how psychological factors could possibly influence physical functioning in this population.

2.4.3 Professional education: increasing psychosocial awareness

The guidelines mentioned previously specify that in order to be defined as a competent specialist for ACHD patient, one needs to know about the psychosocial aspects of adolescence, the transition to adulthood and experience with lifestyle counselling and advocacy ("Care of the Adult with Congenital Heart Disease. Presented at the 32nd BethesdaConference.Bethesda, Maryland, October 2–3, 2000 ", ; Deanfield et al., 2003). A psychologist could be a very useful figure to provide insight and resources when it comes to these areas.

When it comes to professional training, an area which is of the utmost importance is physician-patient communication skills training. The literature on physician-patient communication outlines how many factors affect patient participation in medical encounters, including personal, physician and contextual ones, as outlined by the ecological model (R. L. Street, Jr., Gordon, Ward, Krupat, & Kravitz, 2005). Personal attributes of the patients that effect physician patient communication are their level of education (Beisecker & Beisecker, 1990; R. L. Street, Jr., Voigt, Geyer, Manning, & Swanson, 1995), beliefs about control in the physician-patient relationship (R. L. Street, Jr., Krupat, Bell, Kravitz, & Haidet,

2003), personality (Howell-Koren & Tinsley, 1990), gender (Detmar, Muller, Wever, Schornagel, & Aaronson, 2001; Stewart, 1983), ethnicity (Kim, Klinge, Sharkey, Park, & Cai, 2000; Ward et al., 2003) or confidence in talking to doctors (Kroll et al., 2000).

Certain types of patient behaviours which have been described as being active, such as asking questions, expressing concerns and negative feelings and being assertive can have a powerful influence on the physician's behaviour and decision-making process (R. L. Street, Jr., 2001; R. L. Street, Jr. & Millay, 2001). In fact when patients use these behaviours physicians often respond in a more positive way because they reveal more clearly the patients' needs, beliefs, expectations and preferences (Hines, Moss, & McKenzie, 1997; R. L. Street, 1992; R. L. Street, Jr., 1991)

Also the way the physician communicates can help encourage and discourage patient participation; in particular when physicians use partnership-building and supportive communication patient participation increases (Makoul, 1998; Robinson & Roter, 1999; R. L. Street, 1992; R. L. Street, Jr., 1991; R. L. Street, Jr. et al., 2005; Wissow, Roter, & Wilson, 1994) whereas when physicians enact behaviours which focus on control, patient participation is discouraged (Marvel, Epstein, Flowers, & Beckman, 1999; Waitzkin, 1991). Finally the clinical context, which entails factors such as standards of care, the type of healthcare facility also bear an important influence on patient participation (Bensing, van Dulman, & Tates, 2003; Deveugele et al., 2004; Jones et al., 2004; R. L. Street, Jr., 2003).

There is an indication that patients could be more easily involved if physicians used and supportive communication (R. L. Street, Jr. et al., 2005). Psychologists can play a role in educating the sanitary personnel to communicate effectively, also by presenting strategies to improve physician-patient communication, and to increase patient participation in the health-care process (Kovacs et al., 2006).

Finally, the previously mentioned guidelines specify the importance of ongoing training in sanitary personnel dealing with the ACHD population, and this also includes psychologists. It is important that psychology students, trainees and actual psychologists receive specific training about ACHD issues if they work in this field.

Chapter 3

Psychosocial aspects in ACHD

3.1 Introduction

In order to have a complete understanding of the ACHD patients' life experience it is important to include traditional markers of psychological maladjustment, health related QoL (Kovacs et al., 2006) and also their life experiences. It is also important to investigate how the patients' perceive themselves in their illness and how this perception affects these variables. In this chapter the psychosocial issues in the ACHD patient population will be described, followed by the quantitative research on disease perception, HRQoL and illness behaviour. The life experiences and qualitative studies will be treated with more detail in chapter 5.

3.2 Illness behaviour and disease perception in ACHD

3.2.1 The concept of illness behaviour

It is very important to consider that patients attitude towards their illness, especially in a condition as CHD were an overestimation of the condition could lead to a decreased QoL and an underestimation of it in risk behaviours dangerous for the patients' prognosis. The notion of "illness behaviour" was derived from Parson's concept of "sick role" (Mechanic, 1986, 1992, 1995) and it refers to how individuals interpret and react to their symptoms and also the modalities by which they search for medical advice or cure.

Pilowsky later integrated the sociological concepts of the "sick role" and "illness behaviour", defining illness behaviour as the way in which people react to their psychological and biological functioning (Pilowsky, 1969, 1978a, 1990; Pilowsky, Murrell, & Gordon, 1979; Pilowsky, Spence, & Waddy, 1979). He also identified "abnormal illness behaviour" (AIB) which is defined as dysfunctional

and/or inappropriate perspectives on one's health condition, which persist even when disconfirmed after a medical examination (Pilowsky, 1978a).

There are many different types of dysfunctional health behaviours; as mentioned before inappropriate feelings about one's condition are good examples of this. Often AIB is associated with secondary gain, such as not taking responsibility for one's condition and the refusal of compliance (Winefield, 1991). AIB was investigated in the quantitative study included in this dissertation with the use of the Illness Behaviour Questionnaire (IBQ).

3.2.2 Disease severity rating in ACHD

As stated by Moons et al. (Moons, Van Deyk, De Geest, Gewillig, & Budts, 2005) the conceptualisation of disease severity in CHD is a problematic concept because there are many factors which may have an important influence on the degree of the severity, such as the initial diagnosis, functional status etc. If the conceptualisation of disease severity in the medical field results to be complicated, one can also presume that the patients will find it difficult to give an accurate assessment of the severity of their condition, therefore this needs to be taken into consideration.

In fact, a study carried out in our centre (Chessa et al., 2005) outlined that although the patients had a good grasp of the treatment received, the necessity for follow-up and the prognosis of their condition, they had significant gaps when it came to the anatomy of the heart defect, factors contributing to the onset of endocarditis, the impact of smoking and alcohol, and the possible inheritance of the heart condition.

3.3 Quality of life and ACHD

The impact of disease severity of QoL remains unclear (Kovacs et al., 2005) and the study of the link between illness severity and quality of life is further complicated by the ambiguity surrounding the concept of quality of life itself. In a recent review of 70 articles published in the medical literature, (Moons, Van Deyk, Budts, & De Geest, 2004) noted that the term quality of life was often used inappropriately, probably because it is in vogue (Kinney, 1995) thus drawing conclusions on quality of life when this construct was not really measured.

QoL has been defined as: << the degree of overall life satisfaction that is positively or negatively influenced by individual's perception of certain aspects of life important to them, including matters both related and unrelated to the health >> (Moons, Marquet, Budts, & De Geest, 2004).

In the conceptualisation of QoL specific to ACHD patients, Moons and colleagues have described a "paradigm shift"; instead of investigating quality of life through standardised questionnaires QoL was examined through the exploration of domains in the patients lives, which were both related and unrelated to health. In this study, the most important determinants of an individual's QoL resulted as being: family, job/education, friends, health, and leisure time (Moons, Van Deyk, Marquet et al., 2005)

Very often in the literature there is a comparison between QoL in the Grown up Congenital Heart (GUCH) or ACHD population compared with healthy peers. Some researchers have pointed out that there is a poorer QoL in ACHD patients when compared to normal peers (Kamphuis, Ottenkamp et al., 2002; Lane, Lip, & Millane, 2002; Rose et al., 2005; Simko & McGinnis, 2003) however others have not found this difference (Immer, Althaus, Berdat, Saner, & Carrel, 2005; Saliba et al., 2001).

Most of these studies specify that there appears to be no relation between disease severity or health status and QoL (Kamphuis, Ottenkamp et al., 2002; Rose et al., 2005; Saliba et al., 2001) one study found that the patients who had acyanotic defects had better QoL than those who had single-ventricle physiology (Simko & McGinnis, 2003) whereas the remaining two specify that cyanosis (Lane et al., 2002) and the repair of complete AV-canal (Immer et al., 2005) had a negative impact on QoL,

Other studies investigated the relationship between physical health and objectively measured variables and QoL found no relationship between them (Kamphuis, Ottenkamp et al., 2002; Moons, Van Deyk, De Geest et al., 2005; Ternstedt et al., 2001), although the study by Moons and colleagues did show a minor correlation between functional status and QoL, and also the study by Rose and colleagues mentioned previously outlined that objective cardiopulmonary functioning had a strong impact on cardiac complaints and the physical component of health related QoL (Simko & McGinnis, 2003).

Therefore from all the literature available, it seems that the actual severity of the patients as measured objectively does not have any direct consequences on the

patients' health related QoL. More than actual severity Kovacs et al., (2006) outline that the following variables have been linked with poor QoL in the ACHD population:

1. Some clinical variables, such as cyanosis and orthopaedic problems;
2. Social variables, such as lower levels of educational attainment, age and social impediments;
3. Psychological variables; such as negative thoughts (Fekkes et al., 2001; Rietveld et al., 2002; Saliba et al., 2001).

In the next chapter, the possibility that disease severity as subjectively rated by the patients, as opposed to disease severity as rated by the physician, has an influence on the patients' health related QoL will be outlined. In the quantitative research presented in this dissertation, the relationship between disease severity as rated by the patients and the physicians, and psychological well-being were measured during their hospitalisation before the surgical or catheterisation interventions.

Kovacs (Kovacs et al., 2005) identifies three other areas which affect the QoL of ACHD; ongoing treatment demands, pregnancy and physical activity/exercise.

3.3.1 Ongoing treatment demands

There are various factors which indicate that lifetime medical surveillance is necessary in ACHD patients including increased risk of cardiac arrhythmias, endocarditis and specific risks associated with various forms of CHD (Kovacs et al., 2005). It is not unusual that patients who are initially deemed to be cured will necessitate medical follow-up, therefore it is important to provide facilities which are able to provide long-term medical surveillance and reoperation or further surgery for ACHD (Celermajer & Deanfield, 1991).

As also outlined later on the transition to adult care is not simple as sometimes it is not clear who should take care of this population. In a research on hospitalisation experiences of ACHD patients (Kools et al., 2002), the interviews done with ACHD, their family and the nursing staff on both paediatric intensive care units and adult medical-surgical unit outlined that neither unit is suitable for ACHD patients and that this in turn can create frustration for both health care providers and the patients themselves.

3.3.2 Pregnancy

Most women desire to experience pregnancy and motherhood, however contraception and reproduction issues impose serious concerns for many ACHD women patients. There are contradictions for estrogen pills and intrauterine devices for a series of conditions, and neither gynaecologists more paediatric cardiologists posses the specific skills to address these issues (Somerville, 1998; E. M. Tong et al., 1998).

For women with complex defects pregnancy, labour, and delivery should be carefully monitored and may also be contraindicated in some cases (Somerville, 1998). There might also be anxiety in these women because of the fear of recurrence of CHD in their offspring since the risk is increased, ranging from 3% to 14% (Saidi, Bezold, Altman, Ayres, & Bricker, 1998; Whittemore, Hobbins, & Engle, 1982). These issues can be addressed early in the pregnancy during the second trimester with foetal echocardiography which can verify the presence of significant foetal CHD.

3.3.3 Physical activity/exercise

Exercise has been associated with an improvement in mood in both healthy and unhealthy samples (North, McCullagh, & Tran, 1990) and has also been identified as being a marker of QoL ("Care of the Adult with Congenital Heart Disease. Presented at the 32nd Bethesda Conference. Bethesda, Maryland, October 2–3, 2000," 2001). As outlined earlier, it is possible that patients do not have a clear idea of their condition and that they self-impose physical limitations when it comes to physical activity and in more complex conditions, the opportunity to do exercise is effectively limited. More research is necessary in the area of exercise rehabilitation in ACHD patients ("Care of the Adult with Congenital Heart Disease. Presented at the 32nd Bethesda Conference. Bethesda, Maryland, October 2–3, 2000," 2001).

3.4 Psychological functioning in ACHD

As stated previously, in many respects, having a CHD can be compared to having a chronic condition, because there are various impediments and consequences on the patients' lives and a continuous follow-up which is necessary. One would presume that the severity of the patients' conditions would have an important impact on their psychological wellbeing during hospitalisation and also during recovery, however the literature indicates otherwise.

In a current literature review on the psychosocial aspects related to congenital heart disease (Kovacs et al., 2005) the authors examine the relationship between disease severity and emotional functioning and QoL. When it comes to emotional functioning the authors consider 8 studies, out of which 4 (the ones with the largest samples) suggested that there was no connection between disease severity and psychopathology or emotional functioning (Brandhagen, Feldt, & Williams, 1991; Utens, Verhulst et al., 1998; Utens et al., 1994; van Rijen et al., 2005)

It is interesting to note however that the actual medical condition of the patients and not their rating of their condition did not correlate with emotional functioning, psychopathology and QoL. As shown in the next chapter, it is quite feasible that at least some patients with congenital heart disease either underestimate or overestimate their condition, resulting in a distorted assessment of it, which could lead to different outcomes when it comes to psychopathology, emotional functioning and also quality of life.

Some other qualitative studies on the other hand (Bromberg et al., 2003; Popelova, Slavik, & Skovranek, 2001) outlined that there was a relationship between disease severity and psychological symptoms, and another research suggested that the emotional functioning in CHD patients is poorer than in other populations (Cox, Lewis, Stuart, & Murphy, 2002), however this was not confirmed by another research which compared ACHD patients to reference groups (van Rijen et al., 2003).

When trying to explain why disease severity did not seem to have any effect on psychological functioning, and why it is similar in the ACHD population and reference groups, it was suggested that denial and high achievement motivation could inflate the scores on the self report data (Utens et al., 1994; van Rijen et al., 2003) and there could be significant differences between different cultures (Kovacs et al., 2005). However it could also be that in the qualitative studies the patients are made to be

more aware of their actual condition, bridging the gap between disease severity as rated by the patients and the physician. Another important factor to consider is the limited sample size in these studies; the samples could not have been large enough to demonstrate phenomena of under and over estimation of disease severity.

It is also important to consider the time when the psychological assessment takes place; only the study by Popelova mentioned in the review (Popelova et al., 2001) assesses the patients during the time of referral before hospitalisation. It is interesting to note that depression was associated to older age, worse functional state and unemployment but independent from the severity of cyanosis, the level of the haematocrit, the saturation of oxygen, or previous surgical treatment. Once again, similarly to QoL, functional status seems to be more associated to psychological function than disease severity.

In two more recent studies which assess the psychological state of the patients prior to and after treatment, it also seems that it is not the cardiac condition itself which has an impact on psychological symptoms, but the perception of body image, and the conviction to what degree one can depend on the operated heart that are linked to the patients exhibiting psychological symptoms (Geyer et al., 2006; Norozi, Zoege, Buchhorn, Wessel, & Geyer, 2006).

These studies seem to suggest that the patients focus more on their physical functioning in the here and now, trying to push away elements which can suggest that they are not healthy, such as previous interventions. Perhaps that is why having a physical scar is so distressing; it is physical evidence which reminds the patients that even though now they are well, there could be problems in the future because of their condition.

Another important area relating to the psychological functioning of these patients is body image because the treatment of CHD can entail morphologic differences and surgical scars; this is particularly evident in cyanotic patients. The literature outlines how distortion of the body image, self-consciousness and dissatisfaction with their body is present in this sample and that this could have a negative impact on the patients' wellbeing (Horner et al., 2000; Masi & Brovedani, 1999).

3.5 Social factors in ACHD

Having a CHD most often causes some difficulties when it comes to peer relationships and employment, especially in adolescence and young adulthood, mostly because there is a delay in the maturation progress due to having to cope with the condition, besides the other developmental tasks.

3.5.1 Delayed progression into full adulthood

Parental overprotection is not uncommon in this sample (Brandhagen et al., 1991; L. T. Gantt, 1992) and this might be a factor in delaying the transition of these patients to independence and adulthood. In fact, when compared with health peers, ACHD patients are more likely to maintain dependent lifestyles when compared to their healthy peers (Kokkonen & Paavilainen, 1992) and they seem to develop more knowledge about their condition and assuming more responsibility for their health during late adolescence and early adulthood (Fox, 2002).

3.5.2 Impaired peer relationships

As also outlined in the later chapters, peer relationships are an important resource with coping with CHD especially during adolescence and young adulthood (E. Tong & Sparacino, 1994). Although there are some studies which outline that ACHD patients have adequate outcomes in social functioning (Utens et al., 1994) as also outlined in chapters 5 and 6 adolescents and young adults have a lot of difficulty when it comes to peers, because of physical restrictions which lead to isolation and social awkwardness. The difficulties increase when cyanosis is present (Horner et al., 2000; E. M. Tong et al., 1998).

3.5.3 Employment and insurance

Several studies indicate that ACHD have more difficulty in securing employment and health insurance (Fekkes et al., 2001; Kamphuis, Vogels et al., 2002) and that as a result of physical limitations and problems with schooling they could be more restricted when it comes to employment opportunities (van Rijen et al., 2003).

Unemployment in this population was associated with depression (Popelova et al., 2001) therefore it is important to give adequate vocational counselling when necessary.

Chapter 4

HRQoL and disease perception during hospitalisation in ACHD

4.1 Abstract

Aim of the study: (a) to verify if there is a discrepancy between disease severity rating done by the patients and the physician and also the entity of this discrepancy (b) To investigate the effect of this discrepancy on health related quality of life (HRQoL) and its relation to other psychological factors pertaining to illness behaviour.

Design: Prospective, observational study.

Setting: Patients' admission in a University Hospital.

Patients: 116 patients (52 men 64 women) with a mean age of 38,89 (SD \pm 15,04)

Main outcome measures: disease severity was measured with the use of a numerical rating scale ranging from 0 (least severe condition) – 100 (most severe condition) administered to both patients and physician. Cardiac diagnosis and associated pathologies, previous interventions (both interventional and surgical), general clinical condition and symptomatology current functional status New York Heart Association (NYHA) class and age were utilised when evaluating the patients' severity. QoL was measured with the Psychological General Wellbeing Index (PGWBI) and variables associated to disease perception with the Illness Behaviour Questionnaire (IBQ)

Results: There is a tendency of the patient to overestimate the severity of the disease for low values (< 40) and to underestimate it for higher values. The patients' perception of severity demonstrated a bias of -5.6 (overestimation of the severity of illness), with very high limits of agreement (± 56), confirming the lack of agreement between the two measurements. The distribution of points around the bias is indicative for a better agreement in the low and high severity cases, and a very poor agreement for medium severity cases. When it comes to HRQoL only the patients' perception of severity remained independently associated with the risk of severe distress during hospitalisation and it was also the only independent predictor of the general hypochondriasis subscale.

Conclusion: It is necessary to pay more attention to the patients' perception and knowledge of their condition, and include psychosocial interventions in a multidisciplinary setting aimed to help to bridge the gap between subjective and more objectively rated disease severity and also to provide psychological support when severe distress is present during hospitalisation.

4.2 Introduction

Nowadays more and more patients affected by CHD are reaching adulthood due to the advances and triumphs of cardiovascular medicine and surgery in the 20th century (Gatzoulis et al., 2005). Many of them, depending on the severity of their illness often have to be hospitalised for surgical or catheterisation interventions, therefore it is important to investigate their QoL during these times.

As stated in the previous chapter most studies available in literature outline that there is no relationship between QoL, emotional functioning and disease severity in this population (Brandhagen et al., 1991; Moons, Van Deyk, De Geest et al., 2005; Utens, Bieman et al., 1998; Utens, Verhulst et al., 1998; Utens et al., 1994; van Rijen et al., 2005) although there are other studies which do confirm this relationship (Bromberg et al., 2003; Cox et al., 2002; Kovacs et al., 2008; Popelova et al., 2001); thus it is not yet clear whether the severity of the illness has an impact on QoL and emotional functioning (Kovacs et al., 2005). The study of the link between illness severity and quality of life is further complicated by the ambiguity surrounding the concept of quality of life itself (Kinney, 1995; Moons, Van Deyk et al., 2004).

It is of utmost importance to determine which factors cause distress in this population during the different phases of their condition in order to be able to formulate interventions which aim to improve their QoL and psychological functioning. One of these important phases is hospitalisation previous to an intervention; these are times when the patients could get quite distressed especially if they are uncertain of the outcome, or perhaps because they have never undergone a similar intervention.

Also the conceptualisation and rating of disease severity in CHD results to be problematic since it depends on many factors (such as cardiac malformation, physical functionality etc) and this seems to have repercussions also in the knowledge patients

and relatives have about their condition (Chessa et al., 2005; Moons, Van Deyk, De Geest et al., 2005). Thus it is feasible that there will be discrepancies between the medical and patients' evaluation and perception of disease severity when it comes to CHD. It is therefore important to investigate if this discrepancy actually exists and if it bears an influence in the patients' QoL and psychological functioning.

In this research we set out to investigate the following hypotheses:

1. If there is a discrepancy between disease perception (patients) and evaluation (physician) and also the entity of this discrepancy.
2. The effect of this discrepancy on health related quality of life (HRQoL) and its relation to other psychological factors pertaining to illness behaviour.

This investigation is relevant because the overestimation and the underestimation of the severity of one's condition could both lead to negative outcomes; the condition's overestimation might cause unnecessary distress during hospitalization and its underestimation could lead to health risk behaviours once discharged from hospital. The investigating of the possible presence of a discrepancy between disease evaluation and perception and its effect on HRQoL and psychological functioning during hospitalization in the ACHD population could give very useful indications on the better handling of these patients.

4.3 Patients and methods

4.3.1 Study population

153, Italian speaking patients, aged from 16-73 who were hospitalised to undergo a catheter intervention or cardiac surgeries were asked to participate in this study. Since the objective was to assess the patients' psychological state and QoL before the intervention, the questionnaires were administered during this period.

Patients were excluded if they were assessed as having learning disabilities or genetic syndromes which would not have allowed them to complete the questionnaire. Informed consent was provided orally, and 24 of these patients did not return the questionnaires either because they did not want to participate in the study or because of practical reasons, and 14 returned them incomplete, leaving a total of 116 patients who returned the questionnaires.

Table 4.1. Sample characteristics

Sample age and sex	N° Pts	Marital status	N° Pts
Total final sample: Age-range: 18-73 Mean age: 39.2, SD=14,6.	116 (100%)	Single	56 (48%)
		Married or living with a partner	33 (41%)
		Separated or divorced	7 (8%)
Males	52 (45%)	Widowed	3 (3%)
Patient severity			
Patient severity	N° Pts	Type of intervention	N° Pts
Mild	64 (55%)	Catheterisation	61 (53%)
Significant	27 (23%)	Open heart surgery (OHS)	55 (47%)
Complex	25 (22%)		

4.3.2 Data collection

A. Disease severity perception and evaluation

In order to be able to compare disease severity perception (from the patients' point of view) and disease evaluation (as assessed by the medical physician) we administered a numerical scale to both the medical physician and also to the patients, ranging from 0 the "least possible severity" to 100 the "highest possible severity". The validity and reliability of similar scales have been shown in other studies, where they were also utilised to assess perceived health status in congenital heart disease patients (Badia, Monserrat, Roset, & Herdman, 1999; Moons, 2004; Moons, Van Deyk, De Geest et al., 2005)

The criteria which the medical physician utilised in order to rate the severity of the patients are the following:

- Cardiac diagnosis and associated pathologies;
- Previous interventions (both interventional and surgical);
- General clinical condition and symptomatology;
- NYHA;
- Age.

The rating score was divided into 3 groups of severity by the physician:

1. 0 – 20%: Mild;
2. 21 – 60%: Significant;
3. 61 – 100%: Complex.

B: Psychological General Well Being Index (PGWBI)

The PGWBI questionnaire is a validated HRQoL measure, which has been extensively utilised in clinical trials and epidemiological research in order to provide a general evaluation of self-perceived psychological health and well-being (Boman, Bryman, Halling, & Moller, 2001; Bullinger, Heinisch, Ludwig, & Geier, 1990; Havelund et al., 1999; Hunt & McKenna, 1992; Omvik et al., 1993; Rasmussen, Norholm, & Bech, 1999; Reddy, White, Dunn, Moyna, & Thompson, 2000; Walle, Westergren, Dimenas, Olofsson, & Albrektsen, 1994; Wiklund et al., 1992; Wiklund & Karlberg, 1991) An Italian version of the PGWBI, together with normative data for the Italian population, has recently been made available (Grossi E, 2002).

It consists of 22 self-administered items, rated on a 6-point scale, which aim to assess psychological and general well-being of respondents in six HRQoL domains: *Anxiety* (five items; range 0–25), *Depression* (three items; range 0–15), *Positive well-being* (four items; range 0–20), *Self-control* (three items; range 0–15), *General health* (three items; range 0–15) and *Vitality* (four items; range 0–20). All the questions are referred to the 4-week period prior to the administration of the questionnaire. For each item, there are six response options scored on a 0-to-5 point scale, according to the intensity or frequency of the affective experience. A value of 0 is allocated for the most negative response and 5 for the most positive response. In the following table the range of the scores and the average of the Italian population for each subscore and the total score can be seen.

Table 4.2. PGWBI scores range and mean score for standard Italian population

Dimension	Score	Mean score Italian population
Anxiety	0 – 25 (a lower score indicates a worse state)	17.3
Depression	1 – 15 (a lower score indicates a worse state)	12.4
Positive well-being	0 – 20	11.8
Self-control	0 – 15	11.8
General health	0 – 15	11.1
Vitality	0 – 20	13.4
Total score	1 – 110	78

A total score of “well being” (ranging from 0 – 110 points. 110 being the maximum well being), which is a summary score of all the domains is also obtained. This score has been divided into the following 3 broad categories (Chassany, Dimenäs, Dubois, Wu, & Dupuy, 2004):

1. Positive well-being: 73-110 (this has also been divided into other 2 categories: positive wellbeing: 96-110, non-distress 73-95 (Grossi, Mosconi, Groth, Niero, & Apolone, 2005));
2. Moderate distress: 61-72;
3. Severe distress: 0-60.

C. Illness Behaviour Questionnaire (IBQ)

The notion of “illness behaviour” was derived from Parson’s concept of “sick role” (Mechanic, 1986, 1992, 1995) and it refers to how individuals interpret and react to their symptoms and also the modalities by which they search for medical advice or cure.

Pilowsky later integrated the sociological concepts of the “sick role” and “illness behaviour, defining illness behaviour as the way in which people react to their psychological and biological functioning (Pilowsky, 1969, 1978a, 1990; Pilowsky, Murrell et al., 1979; Pilowsky, Spence et al., 1979). He also identified “abnormal illness behaviour” (AIB) which is defined as dysfunctional and/or inappropriate perspectives on one’s health condition, which persist even when disconfirmed after a medical examination (Pilowsky, 1978a). Often AIB is associated with secondary gain,

such as not taking responsibility for one's condition and the refusal of compliance (Winefield, 1991) and can be measured with the use of the IBQ.

The Italian version of Pilowsky and Spence's IBQ (Fava, Bernardi, Pilowsky, & Spence, 1982; Pierfederici, Bernardi, & Fava, 1982) was used in the study. The questionnaire includes seven scales that evaluate abnormal illness behaviour, which Pilowsky defined as << the persistence of an inappropriate or maladaptive mode of perceiving, evaluating or acting in relation to one's own state of health despite the fact that a doctor has provided a lucid and accurate appraisal of the situation and management to be followed >> (Pilowsky, 1978b, 1993). The instrument is used to assess the patient's feelings toward the significant persons in his or her life, including the treating physicians, and includes questions about the patient's perception of his or her own psychosocial status.

The seven scales of the IBQ are: 1) the general hypochondriasis scale, which assesses the extent of fearful attitudes toward illness that include some insight into the excessive nature of the fear; 2) the disease conviction scale, which measures the strength of the belief that a somatic disorder is present and the degree of reluctance to accept reassurance (questions focused on bodily symptoms and sensations and sleep disturbances); 3) the psychological versus somatic focusing scale, which measures the extent of a psychological versus a somatic focus in perception of the disease (high scores indicate possible adoption of a psychological perspective on the illness; low scores reflect a focus on somatic problems and a tendency to reject the possibility of a psychological dimension to the condition); 4) the affective inhibition scale, on which high scores indicate the inability to communicate feelings, especially negative ones; 5) the affective disturbance scale, which assesses the presence of anxiety, depression, and tension; 6) the denial scale, which measures the tendency to deny life stresses and to attribute all current difficulties to a somatic disorder (high scores indicate a belief that a cure for physical problems would solve all life problems; low scores indicate a belief that ongoing life problems would continue even if the person was physically well); and 7) the irritability scale, on which high scores indicate the presence of interpersonal friction.

Below the table with the range of scores and mean of the Italian hospitalised population can be seen; a score is considered to be pathological if it is greater than the mean + 1 standard deviation.

Table 4.3. IBQ scores range and mean score for standard Italian population

	<u>Mean</u>	<u>SD</u>	<u>Mean + SD</u>	<u>Score Range</u>
General hypochondriasis	3,70	1,99	5,69 (6)	0 – 9
Disease conviction	2,59	1,61	4,20 (5)	0 – 6
Psychological versus somatic focusing	1,77	0,96	2,73 (3)	0 – 5
Affective inhibition	2,67	1,37	4,04 (5)	0 – 5
Affective disturbance	2,47	1,71	4,18 (5)	0 – 5
Denial	2,69	1,21	3,90 (5)	0 – 5
Irritability	2,35	1,51	3,86 (4)	0 – 6

4.4 Procedure

The patients were informed about purpose, procedure and time required of the research by the psychologist during their hospitalisation. After giving oral informed consent they were instructed on how to fill in the questionnaires. Patients were also told that they could change their minds and not fill in the questionnaires at any time, and to give the questionnaires to the head nurse on completion. This study was approved from an independent ethical committee and therefore it has been performed according to the international ethical standards.

One cardiologist scored NYHA, and the NRS (numerical rating scale) of the severity of the condition based on the data from the clinical examination, therefore interrater reliability was not an issue in this study. The cardiologist was also blinded as to the other outcomes measured in the study and the NRS score given by the patient.

4.5 Statistical methods

The relationship between the patient assessed NRS (PTNRS) for severity of illness and the physician assessed NRS (PHNRS) was investigated with an unpaired data Student's t test for means difference, with a linear regression analysis, and with a Bland-Altman analysis. The normal distribution of the PTNRS and PHNRS was tested with a Kolmogorov – Smirnov test with Lilliefors modification. Differences

between PTNRS and PHNRS were explored with a Student's t test or a Mann-Whitney U test depending on the normality of the distribution.

Agreement between the two measurements was tested by calculating the systemic error (bias), and the 95% limits of agreement as the bias \pm 2 standard deviations, as described by Bland and Altman (Bland & Altman, 1986; Bland & Altman, 1999)

The QoL was assessed using the PGWBI and the IBQ. For both the scores obtained from the PGWBI and IBQ the means of all the subscales were compared with the Italian general population scores, both with the whole sample and also with each severity subgroup (both the physician's and the patients' ratings). Both NRS ratings were correlated with the PGWBI subscales (pearson correlation) and total score.

Severe distress (SD) was defined in presence of an overall PGWBI score \leq 60. The univariate association between factors and SD was investigated using an unpaired t test for continuous variables and a Pearson's χ^2 with relative risk analysis for binary variables. Factors being significantly ($P < 0.05$) associated with SD were entered into a multivariable logistic regression analysis. A final model for SD risk was produced, with Hosmer-Lemeshow statistics for calibration of the model.

Discrimination properties of the model were investigated with a receiver operating characteristics (ROC) analysis with the area under the curve (AUC) as accuracy measurement. Adequate cut-off values for SD risk were investigated for sensitivity and specificity.

All data in text and tables are expressed as absolute values with percentage, means and standard deviation; when needed, 95% confidence intervals were calculated.

The means obtained from the IBQ scores were also compared with the ones of other cardiac patients ($N = 192$), including coronary illness patients awaiting a bypass ($N = 122$), myocardium infraction patients ($N = 50$), and patients suspected of having a myocardium infraction who were later excluded ($N = 20$) obtained from the IBQ manual (Fava et al., 1982).

Association between the factors and each single score composing the IBQ was investigated using a correlation matrix for continuous variables and a Student's t test for binary variables. Separate multivariable models (linear or non-linear regression) were settled for each variable composing the IBQ.

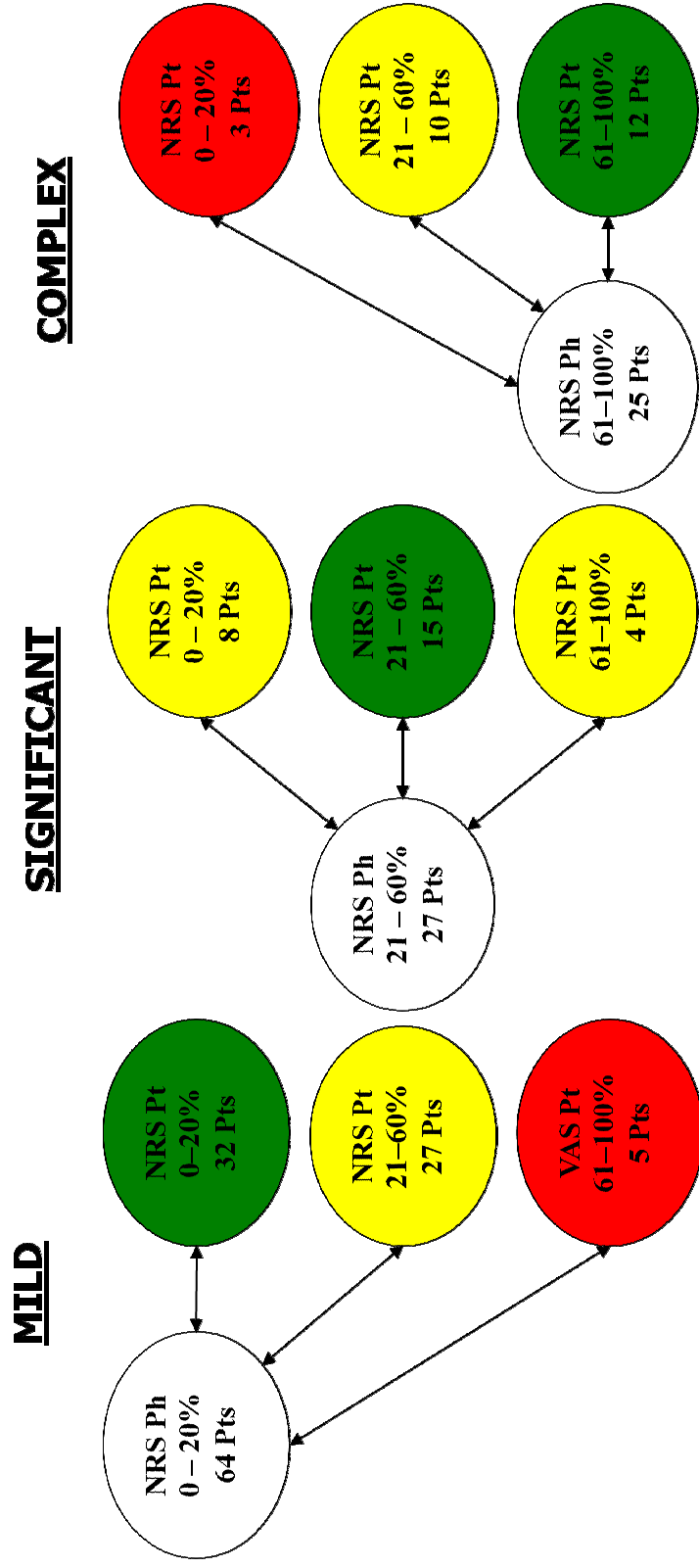
4.6 Results

4.6.1 Patient and physician-assessed severity of illness

In order to get an initial first impression of the correspondence between disease severity evaluation and perception, the three groups of severity as assessed by the physician were compared to the corresponding severity ratings by the patients in figure 4.1.

As can be seen in the diagram, only 51% of the patients had a completely accurate perception of the severity of their condition. Both phenomena of extreme overestimation and underestimation were manifested; out of 64 patients who were evaluated by the physician as having a mild condition, 5 perceived themselves as having a complex one. On the other hand, out of the 25 patients who were evaluated as having a complex condition, 3 felt that their condition was mild.

Figure 4.1. Visual representation of Physician and Patient NRS scores

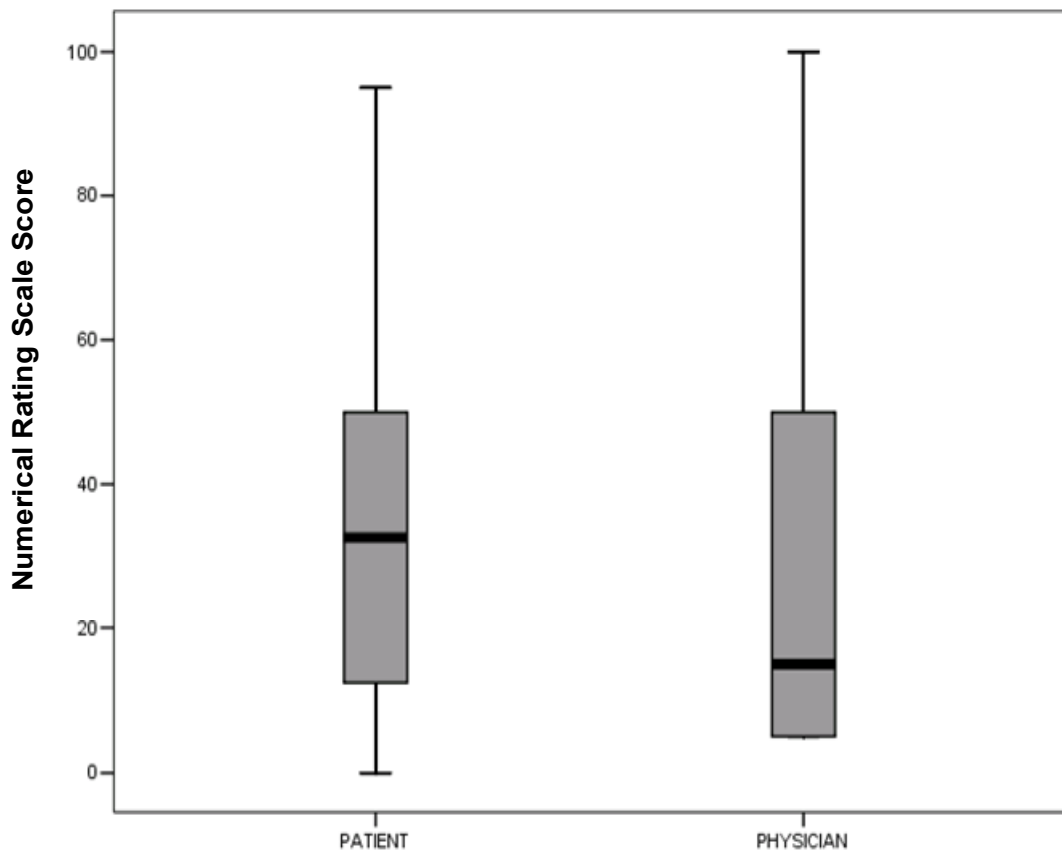


Colour scheme: Green: no overestimation/underestimation, Yellow: mild overestimation/underestimation, Red: severe overestimation/underestimation.

The severity of illness as assessed by the patient with the PTNRS was normally distributed (Kolmogorow-Smirnov Z value = 1.34, p= 0.055) whereas the physician's assessed severity (PHNRS) was not normally distributed (Kolmogorow-Smirnov Z value = 2.59, p< 0.001).

The values of PTNRS and PHNRS are shown in figure 4.2.

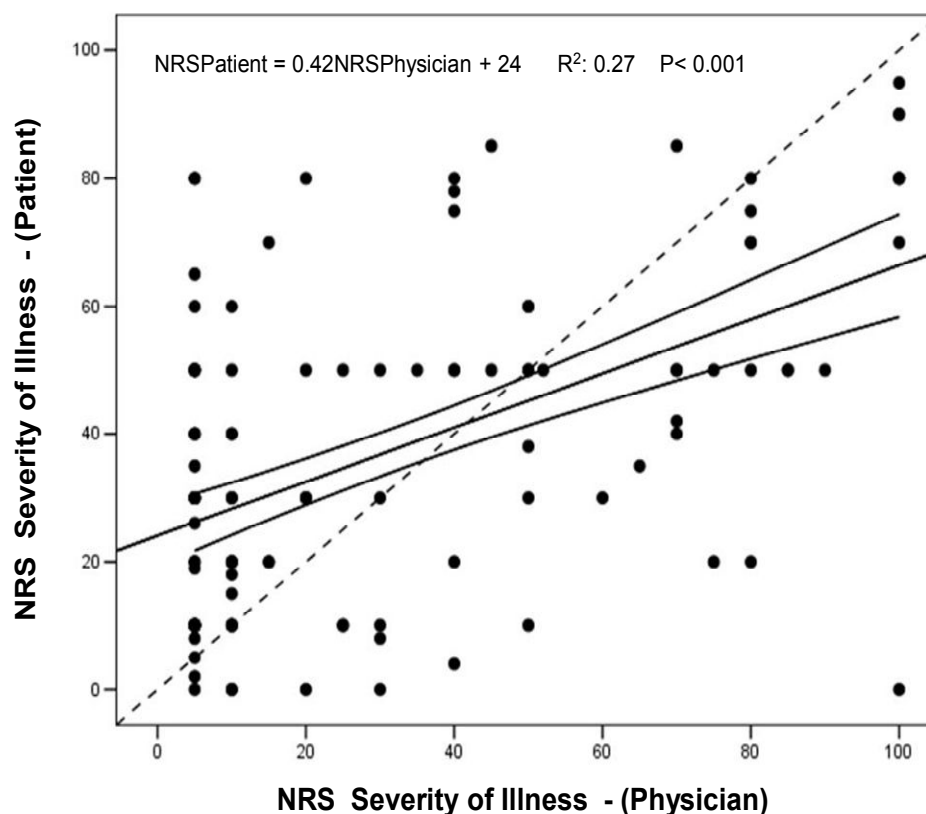
Figure 4.2. Distribution of Patient and Physician NRS Scores



The PTNRS had a mean value of 37.6 ± 25 , median 32.4 (interquartile range 39); the PHNRS had a mean value of 32 ± 31.5 , median 15 (interquartile range 45). When tested for difference, there was a significantly ($p < 0.001$) higher value of the PTNRS at the Mann-Whitney U test.

The two values were tested for association with a linear regression analysis (figure 4.3).

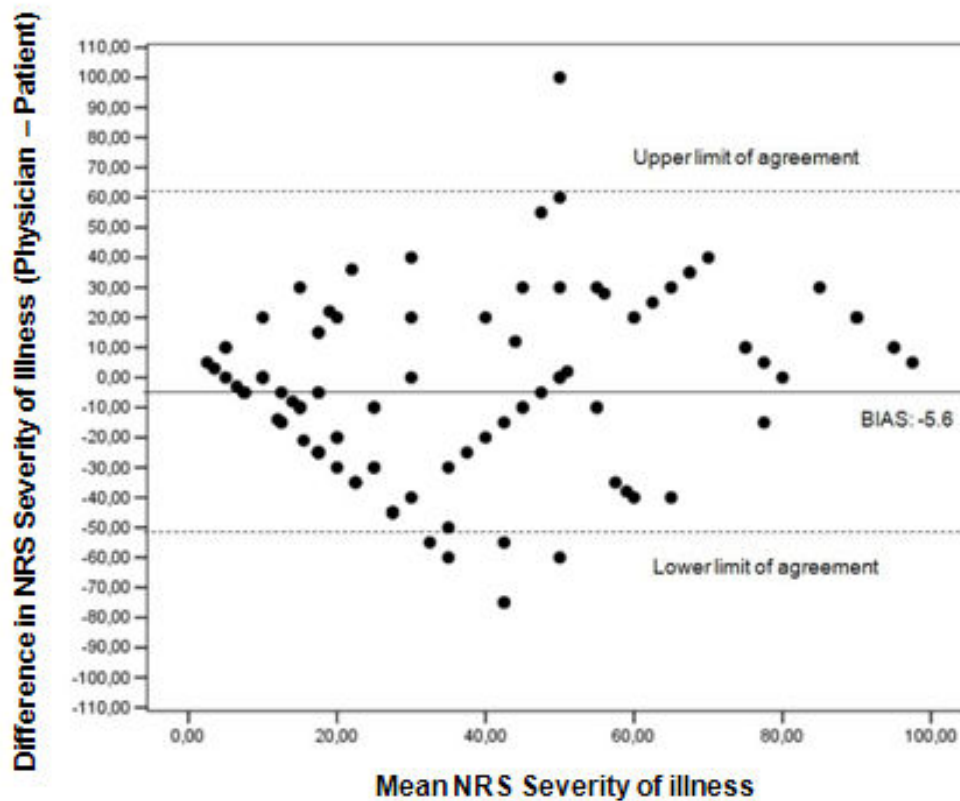
Figure 4.3. Linear regression analysis: Patient and Physician NRS



There was a significant ($p < 0.001$) association. However, the linear regression function is consistently different from the theoretical identity line. In particular, there is a tendency of the patient to overestimate the severity of the disease for low values (< 40) of PHNRS (considered as the reference assessment method) and to underestimate it for higher values of PHNRS.

When tested with the Bland-Altman analysis (figure 4.4), the PTNRS demonstrated a bias of -5.6 (overestimation of the severity of illness), with very high limits of agreement (± 56), confirming the lack of agreement between the two measurements.

Figure 4.4. Bland-Altman Analysis: Patient – Physician NRS



The distribution of points around the bias is indicative for a better agreement in the low and high severity cases, and a very poor agreement for medium severity cases.

4.6.2 Descriptive statistics; QoL and Illness Behaviour

In order to get a first impression of the scores obtained on the questionnaires, in table 4.4 the mean scores obtained in the study population are shown next to the standard Italian population. The study population was then further divided into the 3 main severity categories, as according to the physician and also according to the patients, creating 6 further subgroups.

Table 4.4. PGWBI scores: Italian population, Study sample, Physician and Patient's NRS disease severity subgroups

	Italian pop.		Study Sample		NRS PH 0 – 20%		NRS PT 0 – 20%		NRS PH 21 – 60%		NRS PT 21 – 60%		NRS PH 61 – 100%		NRS PT 61 – 100%	
	Means	DS	Means	DS	Means	DS	Means	DS	Means	DS	Means	DS	Means	DS	Means	DS
Anx	17,3	4,96	17,12	4,75	17,34	4,77	17,49	5,41	16,85	5,04	17,87	3,63	16,84	4,56	14,52	5,11
Dep	12,4	2,62	12,43	2,50	12,58	2,53	12,60	2,41	12,67	2,06	13,04	1,83	11,80	2,86	10,57	3,27
PosWB	11,8	4,02	12,09	3,94	12,25	3,74	12,33	3,90	11,63	3,74	12,38	3,59	12,16	4,71	10,86	4,76
SelfCont	11,8	2,69	11,83	2,85	11,92	2,94	12,65	2,38	11,85	2,76	11,46	2,95	11,56	2,79	11,05	3,15
GHealth	11,1	3,07	10,30	3,07	10,72	2,60	11,02	2,78	10,44	3,24	10,71	2,16	9,08	3,75	7,81	4,25
Vitality	13,4	4,00	13,32	4,03	13,25	4,03	13,88	3,93	13,44	4,45	13,62	3,92	13,36	3,79	11,43	4,15
PGWBITot	78	17,89	77,09	17,73	78,06	17,37	79,98	17,08	76,89	17,72	79,08	15,51	74,80	19,14	66,24	20,74

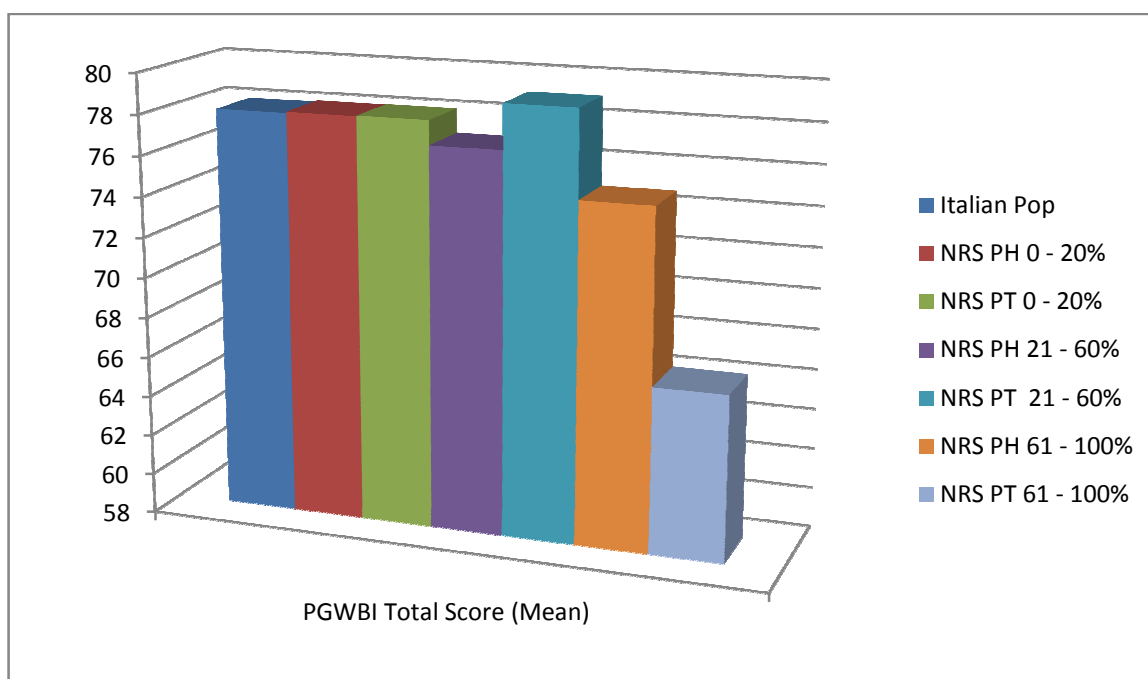
Table 4.5. IBQ scores: Italian population, Study sample, Physician and Patient's NRS disease severity subgroups

	Italian hospitalised pop.		Study Sample		NRS PH 0 – 20%		NRS PT 0 – 20%		NRS PH 21 – 60%		NRS PT 21 – 60%		NRS PH 61 – 100%		NRS PT 61 – 100%	
	Means	DS	Means	DS	Means	DS	Means	DS	Means	DS	Means	DS	Means	DS	Means	DS
Hyp	3,70	1,99	2,24	1,75	1,95	1,57	1,77	1,51	2,41	1,69	2,13	1,53	2,80	2,12	3,48	2,16
DC	2,59	1,61	1,90	1,44	1,81	1,40	1,49	1,32	2,04	1,22	1,81	1,31	1,96	1,74	2,95	1,50
PvsS	1,77	0,96	1,86	0,81	1,83	,83	1,91	,81	2,04	,65	1,92	,79	1,76	,93	1,62	,87
AffInh	2,67	1,37	2,09	1,66	1,92	1,70	1,74	1,61	2,19	1,55	2,38	1,59	2,44	1,69	2,10	1,90
AffDist	2,74	1,71	1,51	1,49	1,67	1,57	1,40	1,55	1,15	1,32	1,44	1,42	1,48	1,42	1,90	1,55
Denial	2,69	1,21	3,71	1,31	3,73	1,21	3,65	1,17	3,78	1,31	3,75	1,31	3,56	1,56	3,71	1,59
Irritability	2,35	1,51	1,66	1,37	1,56	1,27	1,53	1,18	1,67	1,39	1,62	1,37	1,88	1,62	2,00	1,703

A. PGWBI scores: a comparison between sample and standard Italian population mean scores

When comparing the mean scores of the Italian population with the sample mean scores, not many differences emerge. However, when looking at the subgroups, it can be seen that the one with the greatest amount of anxiety, depression and the lowest total HRQoL is the most severe group as evaluated by the patient, as also shown in figure 4.5.

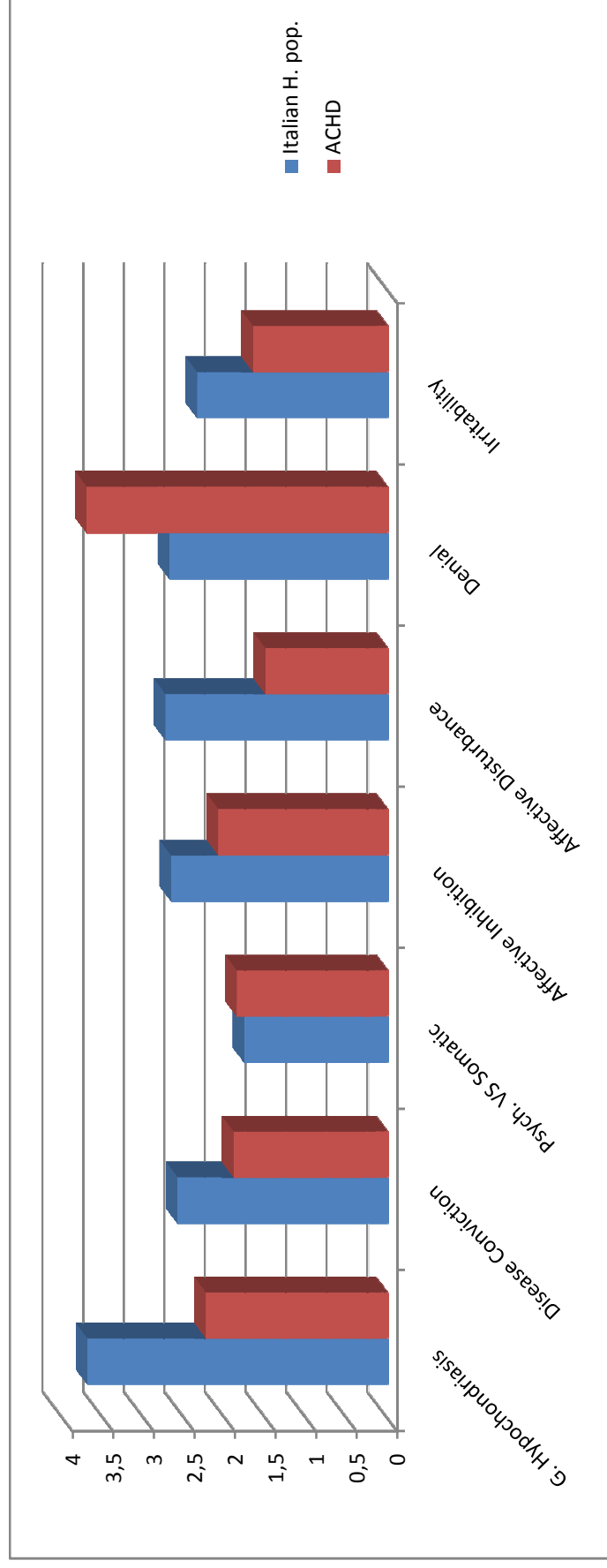
Figure 4.5. Total PGWBI mean scores



B. IBQ scores: a comparison between sample and standard Italian population mean scores

The difference between the mean scores between the sample population and the hospital population can be seen clearly in figure 4.6. Hypochondriasis and affective-disturbance result to be lower and denial higher on average, in the study sample.

Figure 4.6. IBQ mean scores; Italian hospitalised population and study sample



4.6.3 Factors determining the QoL and Illness behaviour

As can be seen from table 4.6, when correlating disease perception and evaluation to the various PGWBI subscales (Pearson Correlation), it can be seen that it is the patients' rating and not the one done by the physician, which correlates with the total score of HRQoL and other dimensions related to it.

Table 4.6. Pearson correlation between NRSPT and NRSPH and PGWBI scores

	Anx	Dep	PosWB	SelfCont	GHealth	Vitality	PGWBITot
NRSPT	-, 207(*)	-, 242(**)	-,128	-,230(**)	-,391(**)	-,219(**)	-,273 (**)
NRSPH	-,049	-,147	-,030	-,049	-,248(**)	-,011	-,094

**<0.01 level (2-tailed)

* <0.05 level (2 tailed)

The QoL was treated as a binary variable according to the PGWBI score. Severe distress (SD) of the QoL was defined as a PGWBI score ≤ 60 . 20 patients (17%) out of 116 presented severe distress during hospitalisation before an intervention. The univariate association between the different factors collected and the presence of a SD was explored (table 4.7).

Table 4.7. Patients' characteristics and association with severe distress

Factor	No severe distress (n=96) Mean \pm SD or number (%)	Severe distress (n=20)	P
Age (years)	38.3 \pm 15.3	41.5 \pm 13.6	0.386
Gender male	45 /47)	7 (35)	0.459
Cyanotic disease	6 (6.39)	5 (25)	0.022
Scheduled open heart operation	44 (45.8)	11 (55)	0.308
Scheduled cath lab procedure	52 (54.2)	9 (45)	0.311
NYHA class	1.68 \pm 0.8	1.9 \pm 0.97	0.278
PTNRS for illness severity	34.1 \pm 23.5	54.3 \pm 29.3	0.001
PHNRS for illness severity	30.3 \pm 30.1	40.2 \pm 37.1	0.200
Previous cath lab procedures (n)	0.67 \pm 1.3	0.601 \pm 1.1	0.838
Previous open heart operations (n)	0.69 \pm 1.03	0.5 \pm 0.82	0.446

NYHA: New York Heart Association; PHNRS: physician visual analogue scale; PTNRS: patient numerical rating scale.

SD was significantly associated with the presence of a cyanotic condition, with the PTNRS, but not with the PHNRS. Cyanotic patients had a SD in 45% of the cases, vs. 14% in acyanotic patients (relative risk 5, 95% confidence interval 1.3-18.4, P). A cyanotic condition affected the PTNRS score (cyanotic: 54.6 ± 31.5 , acyanotic 35.8 ± 24.4 , $P=0.02$).

A multivariable logistic regression analysis including PTNRS and cyanosis was constructed in order to define independent predictors of SD (table 4.8).

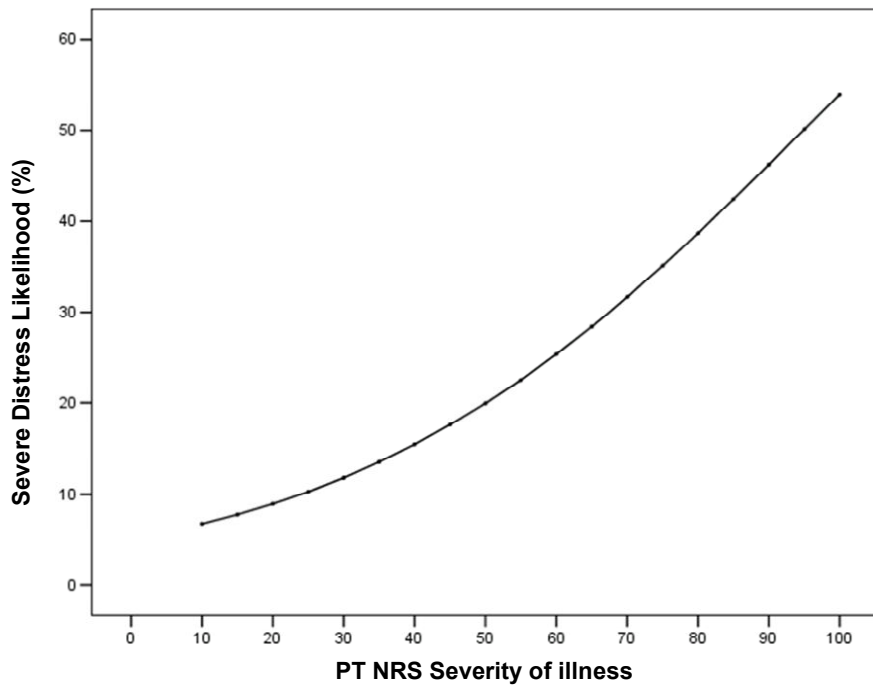
Table 4.8. Multivariable logistic regression analysis for severe distress

Factor	b	S.E.	O.R. (95% C.I.)	P
PTNRS	0.027	0.010	1.028 (1.007-1.049)	0.009
Cyanosis	1.2	0.723	3.306 (0.8-13.6)	0.098
Constant	-2.93	0.569		
Hosmer-Lemeshow χ^2 : 12.6, P= 0.084				

C.I.: confidence interval; OR: odds ratio; PTNRS: patient visual analogue scale; S.E.: standard error.

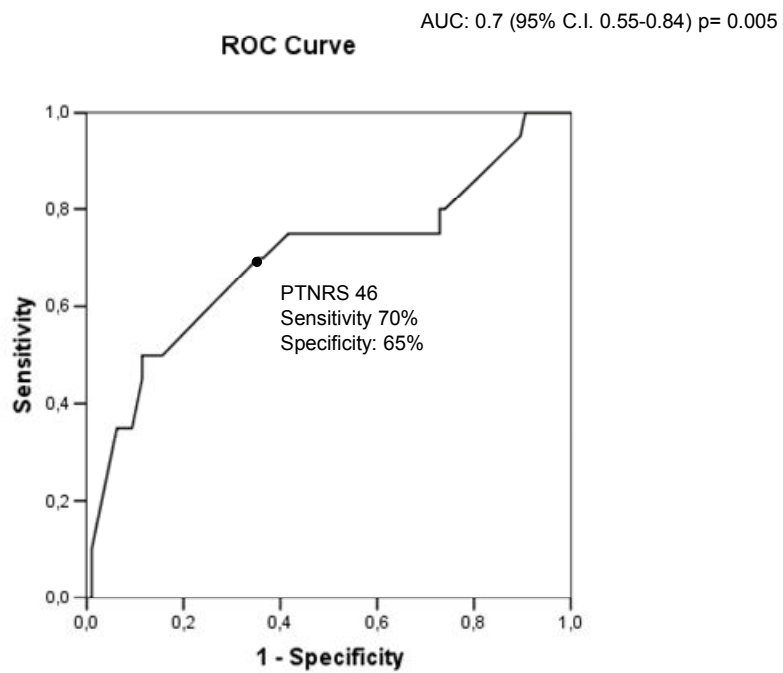
At this analysis, only the PTNRS remained independently associated with the risk of SD. Cyanosis, due to its intercorrelation with PTNRS, was not significantly associated with SD and was therefore excluded from the model. The model based on PTNRS as predictor of SD (figure 4.7) demonstrated a good calibration power at the Hosmer-Lemeshow test (table 4.8).

Figure 4.7. PTNRS as a predictor of SD



To test the accuracy of this model in predicting a SD condition, a ROC analysis was performed (figure 4.8)

Figure 4.8. ROC analysis: PTNRS - SD



The AUC was 0.7 (moderate accuracy, with $P=0.005$). A cut-off value of 46 for the PTNRS had a 70% sensitivity and 65% specificity in predicting a SD. This value confirms that the critical area for PTNRS is in the range of medium severity cases, where the PTNRS - PHNRS relationships intercept the identity line defining the boundary line between over and underestimation of the severity of illness.

Factors determining the Illness Behaviour

Each parameter composing the IBQ was explored for association with the various independent factors considered (table 4.9).

At the univariate analysis, general hypochondriasis score was associated with the NYHA class, the PTNRS, the PHNRS, and the presence of previous open heart operations; the disease conviction score with the PTNRS; the affective inhibition score with the gender (male: 2.48 ± 1.56 , female 1.78 ± 1.68 , $P= 0.024$); the dysphoria score and the irritability score with age. No association was found for the psychosomatic and the denial scores.

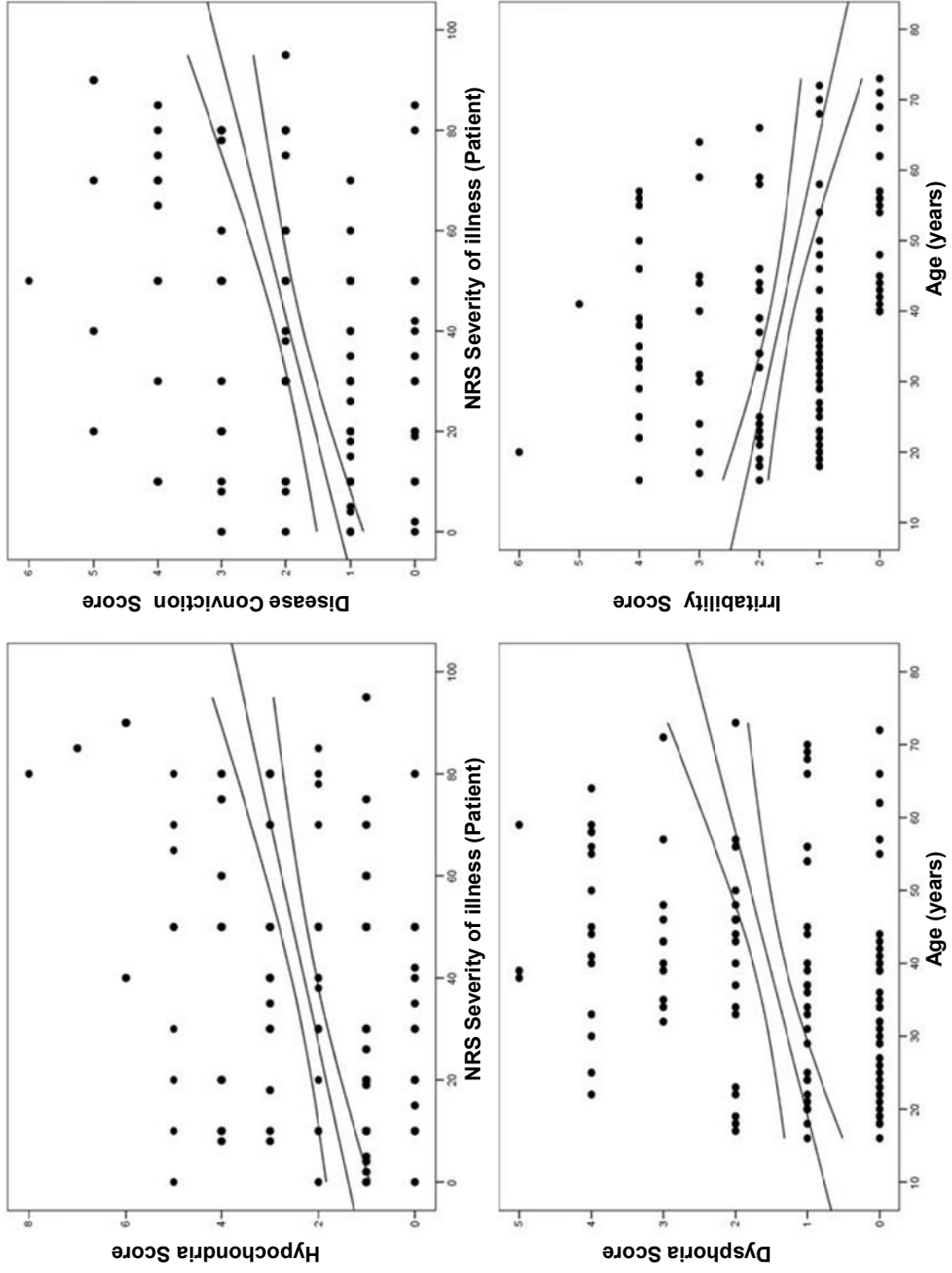
A multivariable analysis was performed for the general hypochondriasis score, and the only independent predictor remained the PTNRS. The association between the various scores composing the IBQ can be seen in figure 4.9.

Table 4.9. Association (P values) between factors and each variable of the IBQ

Factor	General hypochondriasis	Disease conviction	Psychosomatic perception	Affective inhibition	Dysphoria	Denial	Irritability
Age (years)	0.120	0.271	0.316	0.254	0.005*	0.081	0.003*
Gender male	0.493	0.131	0.969	0.024*	0.421	0.914	0.282
Cyanotic disease	0.673	0.258	0.851	0.564	0.932	0.100	0.962
Scheduled intervention	0.094	0.828	0.894	0.098	0.386	0.468	0.079
NYHA class	0.014*	0.411	0.737	0.416	0.928	0.820	0.388
PTNRS for illness severity	0.001*	0.001*	0.233	0.230	0.324	0.607	0.091
PHNRS for illness severity	0.015*	0.343	0.983	0.279	0.320	0.984	0.488
Previous cath lab procedures (n)	0.150	0.497	0.964	0.859	0.263	0.841	0.637
Previous open heart operations (n)	0.049*	0.335	0.330	0.991	0.269	0.381	0.384

NYHA: New York Heart Association; PHNRS: physician visual analogue scale; PTNRS: patient numerical scale; * P<0.05.

Figure 4.9. Association between IBQ scores and the independent factors identified



4.7 Discussion

To our knowledge, this is the first study which made a direct comparison between perceived and evaluated disease severity, comparing these two constructs to HRQoL and psychological constructs pertaining to illness behaviour. As stated by Moons et al. (Moons, Van Deyk, De Geest et al., 2005) the conceptualisation of disease severity in congenital heart disease is a problematic concept because there are many factors which may have an important influence on the degree of the severity, such as the initial diagnosis, functional status etc. If the conceptualisation of disease severity in the medical field results to be complicated, one can also presume that the patients will find it difficult to give an accurate assessment of the severity of their condition, therefore this needs to be taken into consideration.

As suspected there were significant differences between the physician's and the patients' rating of disease severity confirming the literature (Chessa et al., 2005); there was a general tendency of the patients to perceive themselves as being more severe when they were evaluated as having a low severity by the physician (< 40). Both underestimation and overestimation occurred and there was a very poor agreement for medium severity cases.

Therefore it seems that there is a substantial amount of patients who are not aware of their real condition during hospitalization and that possibly psychological factors distort their perception. As stated previously, the literature indicates that the impact of disease severity of QoL remains unclear (Kovacs et al., 2005) and the study of the link between illness severity and quality of life is further complicated by the ambiguity surrounding the concept of quality of life itself (Kinney, 1995; Moons, Van Deyk et al., 2004).

As stated in the previous chapter, the literature tends to indicate that objective health does not affect quality of life and psychological functioning. This study confirms the results obtained from the literature; the physician assessed severity does not seem to influence or be related to HRQoL or psychological functioning and it also gives a possible reason why there seems to be no relationship between these variables; the patients seem to have a distorted perception of their severity. Some patients, especially those with a severity evaluated to be around 50 by the physician, seem to underestimate or overestimate their condition, resulting in a distorted perception of it,

which appears lead to different outcomes when it comes to psychopathology, emotional functioning and also quality of life.

It also outlines that disease severity as rated by the patients is an important predictor when it comes to the patients' well-being and psychological state; in fact, in this study the patients' rating of severity resulted as being the only independent predictor of severe distress. The patients' rating of severity also resulted as being the only predictor of the subscale "general hypochondriasis" from the IBQ questionnaire; this indicates that an excessive fear of being ill results in perceiving oneself as being more seriously ill.

These variables could also be influencing each other; for example anxiety and depression could distort one's perception and perceive themselves as being more serious than they actually are, and also an incorrect evaluation of one's condition due to education and other cognitive factors could give rise to an incorrect evaluation leading to a decreased HRQoL.

4.8 Clinical Implications

It is important to pay further attention to what the patients know about their condition, because it seems that it can have an impact on their QoL during their hospitalisation. Psychological support should be offered to patients suffering from severe distress and when this is associated to an overestimation of the severity of the condition the patient should be reassured. In the cases of underestimation of the severity of the condition it would be advisable to counsel the patient after the procedure, in order to safeguard their health and to try to improve therapeutic adherence.

4.9 Limitations

Since in practice there is no standard for the classification of the severity of congenital heart defects (Moons, Van Deyk, De Geest et al., 2005), especially one which can be compared to the patients' rating, we utilised a NRS for both physician and patients. Although the physician took into account specific criteria when formulating her judgement, these have not been quantified and validated. However,

when correlating this score to previous intervention and NYHA a high correlation was obtained, confirming the proper use of at least these criteria.

Another limitation of the current work is the size of the sample. Before the results can be generalised the sample should be increased further, even though the sample is quite representative when it comes to the percentage of patients fitting into the severity groups: mild (55%), significant (23%) and complex (21%).

Finally the instruments utilised were targeted for an Italian population and also an Italian hospitalised population, but since no specific CHD instrument (such as the congenital heart Disease - TNO/AZL adult quality of life) has been validated in Italy they could not be utilised for this study, perhaps missing out on some important issues specific to this population, which hopefully have been dealt with in the qualitative research presented in this dissertation.

4.10 Conclusions

This study outlines that there is a significant discrepancy between disease severity as rated by the patients and as assessed by physicians in ACHD. Moreover, the patients' severity rating influences HRQoL, with highly rated severity (also when overestimated) being the only variable which increases the probability of severe distress during hospitalisation, and which is also associated to general hypochondriasis. It is therefore necessary to pay more attention to the patients' perception and knowledge of their condition, and include psychosocial interventions in a multidisciplinary setting aimed to help to bridge the gap between disease severity as rated by the patients and the physicians and also to provide psychological support when severe distress is present during hospitalisation.

Chapter 5

Life experiences and coping with CHD

5.1 Chronic illness, disability and coping: an Integrative Conceptual Framework

Chronic illness has been defined as << an illness that persists for a long period of time... lasting 3 months or more >> by the definition of the U.S. National Centre for Health Statistics. (MedicineNet, 2008).

Burish e Bradley (Burish & Bradley, 1983) make a distinction between acute and chronic illness based on four dimensions of the illness:

1. **The cause:** it is usually infective for acute illness and it depends on lifestyle factors with it comes to chronic illness and disability (CID);
2. **Time-line:** brief when it comes to acute illness and chronic when it comes to CID;
3. **Identity:** the individual usually has a clear idea of the symptoms and the causes connected to the condition whereas in chronic illness the symptoms usually manifest themselves at an advanced stage and the condition might not have a single specific cause;
4. **Outcomes:** acute illness is usually cured over time with the proper treatment, whereas chronic illnesses continue to persist for the rest of a person's life despite treatment.

Sperry (Sperry, 2006) has identified four types of chronic illnesses:

1. Life-threatening diseases
 2. Non-life-threatening chronic conditions, which are often manageable;
 3. Progressively sever diseases and;
 4. Diseases that fluctuate in symptoms, but are not life-threatening;
- specifying that each type tends to require different types of biopsychosocial counselling and therapy.

According to the most recent definition of Disability provided by WHO, disability implies s form of activity limitation or restriction in participation (*World*

Health Organization 2008) and the definition give by the US's Social Security Administration (*Social Security Administrator*, 2008) specifies that the mental or physical impairment which does not allow to do any substantial work has to last for at least one year and result in the individual's death.

When it comes to CHD, since there is such a wide spectrum of severity some conditions could be classified as being acute, because they are resolved by an intervention, others as being chronic, since at the moment no surgical or procedural intervention which can resolve the condition is available. A percentage of these patients, the most severe ones, can be considered as being disabled, because they are extremely limited in many life spheres and are unable to work.

Livneh and Martz (Livneh & Martz, 2007) have identified the following elements when examining the definitions of coping given by various authors:

1. Coping is a broader construct than defending;
2. It is sometimes seen as a global personality trait but it is in no way meant to be inflexible, trans-situational psychological construct;
3. Coping efforts include a wide range of cognitive, emotional and behavioural strategies which are directed at both external stressors and internal demands;
4. Coping efforts (both stable and process-oriented aspects) should not be confused with psychosocial outcomes.

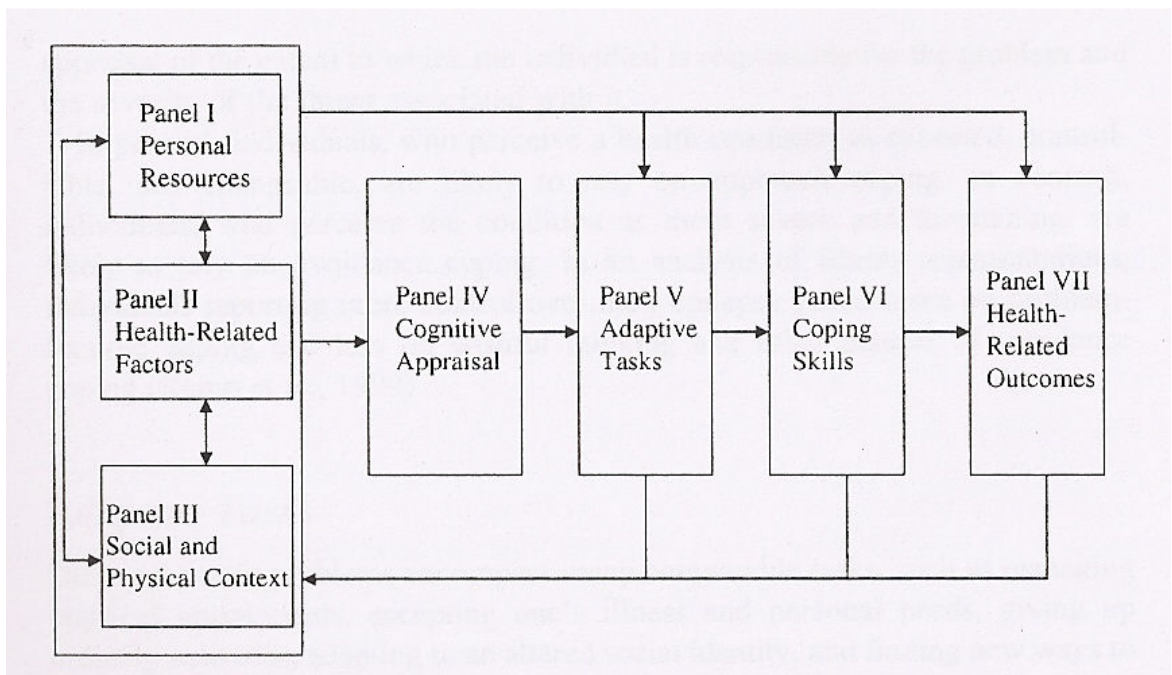
These authors also made the following considerations, on coping based on several review papers (R. S. Lazarus, 1993; Richard S. Lazarus & Folkman, 1984; Moos & Schaefer, 1984; Zeidner & Endler, 1996) which are useful when trying to evaluate the effectiveness of coping:

1. Coping strategies vary in time and between individuals;
2. Their effectiveness depends on the type of stressor, severity, duration and context of the experienced stress;
3. A good balance of both person and environmental strategies are necessary in order to manage the stressful situation;
4. For coping to be adaptive, a versatile repertoire of coping strategies including both problem-focused and emotion-focused efforts should be available, according to the circumstances;
5. Finally all coping strategies may be viewed as a mediating factor between stressful encounters and the final psychosocial outcomes, regardless of their level of effectiveness.

Many authors have specified that coping efforts to overcome functional limitation linked to chronic illness and disability (CID) can be seen as one of the cardinal links between factors related to the condition in itself, such as severity, duration, time and manner of onset and quality of life (Livneh, 2001; Maes, Livneh, & de Ridder, 1996; Moos & Schaefer, 1984).

Moos and Holohan (Moos & Holahan, 2007) propose an Integrative Conceptual Framework to guide the process of coping with CID, consistent with other conceptual models of adaptation to health problems (Livneh & Antonak, 1997; Livneh & Parker, 2005; Walker, Jackson, & Littlejohn, 2004).

Figure 5.1. The conceptual model of the determinants of health-related outcomes in CID. Moos & Holohan (2007) (pg 110)



This model collocates coping skills in a broader predictive model; five sets of factors are associated with the selection and choice of Coping Skills (Panel VI):

- **Personal resources** (Panel I): include factors such as stable personality traits considered being positive, intellectual ability , prior coping-experiences and demographic variables.
- **Health related factors** (Panel II): include disease characteristics such as onset, stage, symptoms and severity and the health-care environment and treatment procedures.

- **Social and Physical context** (Panel II): encompasses relations between patients and their social support networks, including their family members and friends and also the caregivers and co-workers they are in contact with.
- **Cognitive Appraisal** (Panel IV): this is the way in which individuals appraise a crisis and it includes the context of the stressor, how much it was expected and if there was enough time to prepare for it. It also includes the perception of whether the condition is a challenge or a threat and how much one is to blame for it.

Adaptive tasks (Panel V): the authors identify seven main tasks divided into two groups:

1. Tasks related to the health condition and its treatment
 1. Managing symptoms
 2. Managing treatment
 3. Forming relationships with Health-care Providers
2. General tasks which apply to all types of life-crises.
 1. Managing emotions
 2. Maintaining a Positive Self-Image
 3. Relating to Family Members and Friends
 4. Preparing for an Uncertain Future

Finally the choice of **coping skills** (Panel VI) then mediates with the influences of Panels I through V and **health related outcomes** (Panel VII) in a mutual feedback cycle creating more long-term health outcomes.

5.2 Approach and avoidance coping skills

When it comes to the categorisation and classification utilised in this conceptual framework, the two approaches which have been previously utilised in the literature (the focus of coping and the method of coping) to classify coping strategies have been combined in four broad domains, each including two subsets which can be seen in this table:

Table 5.1 Approach and avoidance coping skills

	Approach	Avoidance
Cognitive	<p>Cognitive approach coping</p> <ul style="list-style-type: none"> • Logical analysis and the search of meaning • Positive reappraisal 	<p>Cognitive avoidance coping</p> <ul style="list-style-type: none"> • Cognitive avoidance or denial • Acceptance or resignation
Behavioural	<p>Behavioural approach</p> <ul style="list-style-type: none"> • Seeking guidance and support • Taking problem-solving action 	<p>Behavioural avoidance coping</p> <ul style="list-style-type: none"> • Seeking alternative rewards • Emotional discharge

5.2.1 Approach coping strategies:

- **Logical analysis and searching for meaning:** this includes trying to break down a crisis into manageable parts, and using cognitive efforts to learn more about the condition to handle it better. This category also includes those attempts to give meaning to the course of events in order to anticipate the grieving process;
- **Positive reappraisal:** it includes the acceptance of the basic reality of the situation but also restructuring it in a more positive light;
- **Seeking Guidance and support:** include efforts in obtaining information about health conditions and treatment and procedures options and their probable outcomes. Seeking emotional reassurance from family, friends and caregivers, and also religious coping behaviour are included in this category;
- **Taking Problem-Solving Action:** taking action to deal with situations in a direct way and it includes how to control symptoms and reorganising the environment to adjust to the condition.

5.2.2 Avoidance coping strategies

- **Cognitive Avoidance or Denial:** responses aimed at denying or minimising the seriousness of a crises, which can temporarily protect an individual from feeling emotionally overwhelmed and allows him/her to gather the necessary resources to deal with the situation.
- **Acceptance or resignation:** coming to terms with the reality of a health condition and acknowledging the impossibility of changing things. It is especially helpful when approaching death, freeing the individual with CID from distress and helping them to find a deeper meaning.
- **Seeking alternative rewards:** attempting to replace the losses endured because of the health condition by changing one's activities and creating new ways of obtaining satisfaction.
- **Emotional discharge:** Openly venting out distressing feelings, using jokes and humour to try to deal with distress. It also entails "acting out" by not complying with a treatment regimen and behaviours which might temporarily reduce tension which may lead to a temporary failure of affective regulation, as individuals alternate between emotional control and emotional discharge.

5.3 Coping strategies and health related outcomes

Wright warns about the many possible biases when it comes to the assessment of coping strategies (B.A. Wright, 1983; B.A. Wright, 1991, 2007) therefore it is important to consider all the factors mentioned previously in the Integrative Conceptual Framework, in order to make an assessment of how adaptive certain coping strategies are. However, although coping strategies cannot be judged as being more functional or dysfunctional a priori, there are some studies which link certain types of strategies with health-related outcomes.

In fact the literature indicates that generally approach-coping strategies are usually associated with fewer symptoms, less distress and more well-being (Horgan & MacLachlan, 2004; Livneh & Antonak, 1997; Penley, Tomaka, & Wiebe, 2002) even though there are some important exceptions.

The most findings are most clear when it comes to the following approach coping strategies; positive reappraisal, seeking guidance and support and problem

solving. Often individuals become more resilient and achieve personal growth and a wider range of coping skills as a consequence of having a CID. The approach-coping skills of positive reappraisal and taking problem-solving action are essential in supporting this growth (Sheikh, 2004; Urcuyo, Boyers, Carver, & Antoni, 2005; Widows, Jacobsen, Booth-Jones, & Fields, 2005). On the other hand, avoidance coping strategies, especially cognitive avoidance and emotional discharge, tend to be associated with poorer psychological outcomes (Moos & Holahan, 2007).

Also avoidance coping can affect health-related outcomes (Roesch et al., 2005) and the utilisation of behavioural disengagement and resignation has been associated to a higher likelihood of mortality from chronic heart failure (Murberg & Bru, 2001). One of the main reasons for avoidance coping being linked to poorer health outcomes is its link to lack of adherence to treatment recommendations (Sherbourne, Hays, Ordway, DiMatteo, & Kravitz, 1992).

Cognitive avoidance may also have temporary benefits, especially when it is used in early stages of a traumatic event (Goldbeck, 1997; Lustig, 2005). Denial may also be beneficial in dealing with aspects of health problems that are unalterable although it could hinder the re-appraisal of relationship or employment issues (Pakenham, Stewart, & Rogers, 1997) Emotional blunting may also be of benefit if the treatment is accepted and it is necessary to undergo necessary but difficult procedures (Heim, Valach, & Schaffner, 1997).

Sometimes approach coping is also not appropriate and can prove to be detrimental; for example if patients try to gain active control in situations which cannot be changed, feelings of undue personal responsibility, blame and guilt can ensue (Astin et al., 1999). Also more passive coping styles were associated with more favourable adherence amongst individuals receiving hospital-based, provider-controlled treatment (Christensen, 2000).

In conclusion, the gains obtained from the coping strategies also depend on the other resources; in a research on coping strategies and cardiac patients utilising the Integrative Conceptual Framework outlined in this chapter, positive aspects of relationships enhanced coping efforts which were linked to psychological adjustment. On the negative aspects of relationships had a bad effect on coping strategies and were linked to poorer psychological adjustment (Holahan, Moos, Holahan, & Brennan, 1997)

5.4 Life experiences of ACHD patients and coping with CHD

As stated previously, when it comes to disease severity, the ACHD population is a highly heterogeneous one and this entails very different life experiences (Verstappen et al., 2006). Those patients who are born with a complex 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 their everyday lives (Moons et al., 2002). As already mentioned, ACHD encounter specific psychosocial challenges which may have an impact on their emotional functioning, self-perception, and peer relationship (Kovacs et al., 2005).

Having said this, no specific psychosocial outcomes referring to patients born with the same pathology have been found; patients having the same conditions can have widely varied experiences depending on many biopsychosocial variables. However there appear to be some patterns of experience and perspective in this broad diversity (Verstappen et al., 2006). Understanding the life experiences of adults with congenital heart disease can help discover those factors which helped and hindered them in their growth. Additionally they can give us precious insight on how to improve the patients' stay during hospitalization. Another final important contribution is that they can help us improve the patients' compliance and evaluate if they are utilizing maladaptive coping strategies which could lead to a negative prognosis.

There are currently very few studies which take into consideration the direct experiences of ACHD patients themselves, even though their direct experiences are crucial in order to be able to adequately counsel these patients and their families in order to promote quality of life (Claessens et al., 2005). There have been two studies conducted on the adolescent population (McMurray et al., 2001; E. M. Tong et al., 1998).

Both these studies outline how adolescents with CHD struggle with physical limitations and face social exclusion. The limitations of the patients depended on the severity of the condition and often the parents resulted as being overprotective towards them, in fact the literature indicates that approximately one fourth of ACHD patients recall their parents being overprotective during their childhoods and adolescence (Arnett, 2000; Brandhagen et al., 1991; McMurray et al., 2001). This could explain why in general these patients tend to live longer with their parents as over protection could lead to less autonomy (Gersony et al., 1993; Kokkonen &

Paavilainen, 1992). In both researches the theme of being and the attempts to cope with this emerged strongly.

The theme of being different from healthy peers also emerges in the literature concerning adults are followed by attempts and struggle to feel normal and to be perceived as being normal by others (Berghammer, Dellborg, & Ekman, 2006; Claessens et al., 2005; L. Gantt, 2002; L. T. Gantt, 1992; Verstappen et al., 2006) and this can be followed by feelings of ambivalence and also denial of the condition and efforts to exceed their physical boundaries (Berghammer et al., 2006).

In one study there was a focus on the problems encountered by the female patients (L. T. Gantt, 1992) with CHD, mainly the concerns regarding fertility, contraception, and pregnancy. The patients also showed concern about their surgical scars and body size and were uninformed about aspects regarding their condition and reproductive matters. It seems that women experience a negative body image as a consequence of scarring and cyanosis more than men, who tend to experience a negative body image only during adolescence (Claessens et al., 2005)

In one other study (Kools et al., 2002), the hospitalisation experience of ACHD patients were examined, by taking into consideration multiple perspectives; including also the ones of the patients families and the nurses during acute phases of their illness. The authors highlight the fact that each groups had different expectation about the management of the physical needs and about the way the patients and the patients' families behaved and that this caused interpersonal conflict, distrust, anxiety and dissatisfaction with the health-care provided.

The patients report on how equipment appropriate for adults was not always available or considered, that they were sometimes treated in a condescending way, scared of unfamiliar treatment protocols, and they felt that they were not listened to when they tried to make some observations about their health (Kools et al., 2002).

In one article the authors report quotations from the Adult Congenital Heart Association in order to address the patients' perspectives (Verstappen et al., 2006) in order to outline the implications for care. The following section is a summary of the most important issues which emerged combined with other literature.

Patients being told they were "miracle babies" and also at the other end, lost causes, that they would not survive for long, erroneously. When the patients believed that they were lost causes they sometimes engaged in highly risky behaviour because

they did not believe they had a future to put at risk, and they also had difficulty to move into adulthood since they were not expecting to make it so far.

The risk of language based misperception in understanding if the condition is completely cured or not in the conditions of moderate and great complexity. Few patients have the acknowledge and understanding to enact behaviours to safeguard and optimize their health (Chessa et al., 2005; Moons et al., 2001) and this could also occur because some health care providers have attitudinal barriers to educate the patients on risks associated to the condition and the need for life-long follow-up. Also the patients' behaviours can contribute to a lack of knowledge as outlined previously.

Adult patients also commonly report that they have difficulties when they have to go to the "regular" medical system, where they sanitary personnel is not specialised in CHD. They often report the need to be vigilant in order to reject medications which could be inappropriate for them, prescribed by the primary care providers and that insisting for special care can result in hostility.

Being an invisible disability, ACHD patients often report that their family and friends imply that the problems they have do not really exist and that changes in work or household responsibilities are not really necessary. These patients might also have problems at work as their requests can be met with scepticism or accusations of dishonesty.

Other difficult situations the patients have to face are making difficult decisions regarding treatment, referral for surgery and being informed that there is nothing left to be done (Horner et al., 2000). Sometimes information is withheld from the patients and they get to know about their real condition at the onset of new problems, making it particularly hard for them to adapt, also because of feelings of betrayal and anger.

Finally it is also important to emphasise that there are not only negative consequences but also benefits that patients derive from surviving a CHD condition; these include treats from their family because they had to endure things, receiving special attention, increased resilience and maturity and a clearer sense of their vocations. The on-going awareness of one's mortality can lead to an increased appreciation of life, more clarity of purpose and better decision making (Mathieu, 2005).

Chapter 6

Life experiences and coping strategies in ACHD

6.1 Abstract

Background: The population of ACHD is continuously rising, and the patients have to face many psychosocial concerns due to their condition.

Aim: The aim of this study was to explore the experiences of ACHD patients hospitalised in our centre, from when they become aware of their experience, till after the open heart surgery they underwent. Successively, the patients' perception of their condition was also taken into consideration in the final discussion, uniting both quantitative and qualitative data.

Methods: Unstructured, in-depth interviews were performed with 11 patients, ranging from 20 to 56 years, who were hospitalised to undergo open heart surgery within the hospital. Interviews were recorded, transcribed and analysed according to Grounded Theory procedures.

Results: In accordance to the literature available, the patients being different to others who are healthy resulted to be the core category which emerged. When the patients' perception of the severity of their condition was highly discordant, this resulted in the adoption of coping strategies which did not seem to be adaptive and could have put the patients' health at risk.

Conclusion: It is important to help the patients elaborate an accurate perception of their condition and to utilise the resources they possess in an optimal way, in order to increase for them to cope with their condition in the best way possible.

6.2 Introduction

Even though , the ACHD population is a highly heterogeneous one and this entails very different life experiences (Verstappen et al., 2006), in many respects, having a CHD of a certain severity can be compared to having a chronic condition, because there are

various impediments and consequences on the patients' lives and a continuous follow-up which is necessary (Moons et al., 2002). The psychosocial challenges which these patients have to face have already been outlined in the previous chapters; the most important elements to take into consideration are that the patients encounter specific psychosocial challenges which may have an impact on their QoL, emotional functioning, self-perception, and peer relationships (Kovacs et al., 2005). In the qualitative research present in the literature the theme of being different from healthy peers also emerges very strongly (Berghammer et al., 2006; Claessens et al., 2005; L. Gantt, 2002; L. T. Gantt, 1992; Verstappen et al., 2006).

6.3 Method and sampling

The method of Grounded Theory was utilised to analyse the data; it was first developed by two sociologists, Glaser and Straus, during their collaboration in a research on hospitalised patients who were dying (B. G. Glaser & Strauss, 1965). They defined it as being the general method of comparative analysis, and a set of procedures capable to systematically generate a theory based on data (B. G. Glaser & Strauss, 1967). Since their first publications, there was a split in theory between them which was consolidated by the publication of Strauss of several works (Strauss, 1987; Strauss & Corbin, 1990), followed by a rebuke of Glaser (B. Glaser, 1992).

Successively a constructivist approach was developed by Charmaz; she makes a distinction between objectivist and constructivist concepts of Grounded Theory; the former takes for granted the presence of a neutral observer whereas the latter << recognizes that the viewer creates the data and ensuing analysis through interaction with the viewed >> (K. Charmaz, 2000) p.523 and this approach was utilised in the analysis of the data (K. Charmaz, 2006).

A particular type of sampling called theoretical sampling is used in Grounded Theory; the selection of the sample is linked to the analysis presuppositions and the procedures of the method. As opposed to probabilistic sampling where the researcher's goal is to representatively capture all the possible variations in a sample, in this case the objective is to gain a deeper understanding of the analysed cases in order to facilitate the development of concepts and categories and finally a theory.

A single, unstructured, in-depth research was conducted with ACHD patients who were hospitalised in our centre and who were due to have open heart surgery. The

interview took place after the intervention, once the patient was fully recovered. Approval was obtained from the Ethical committee of reference. The focus of the interviews was their previous life experiences, including the quality of their stay in hospital, in order to be able to develop psychosocial interventions aimed to support the patients which necessary. 11 patients were interviewed; the number of the subjects selected is decided on the basis of saturation as specified by Grounded Theory procedures.

Since these patients also filled in the quantitative questionnaires, their evaluation of the severity of the severity of their condition was incorporated in the discussions. Below is a table of the patients in the order they were interviewed. The total sample consisted of 7 males and 4 females, ranging from 20 to 56 years. Most patients had a complex condition and had previously undergone either catheterisation or open heart surgeries. Only one patient (3) had an initial heart defect which was of a cyanotic nature.

Table 6.1. Sample characteristics

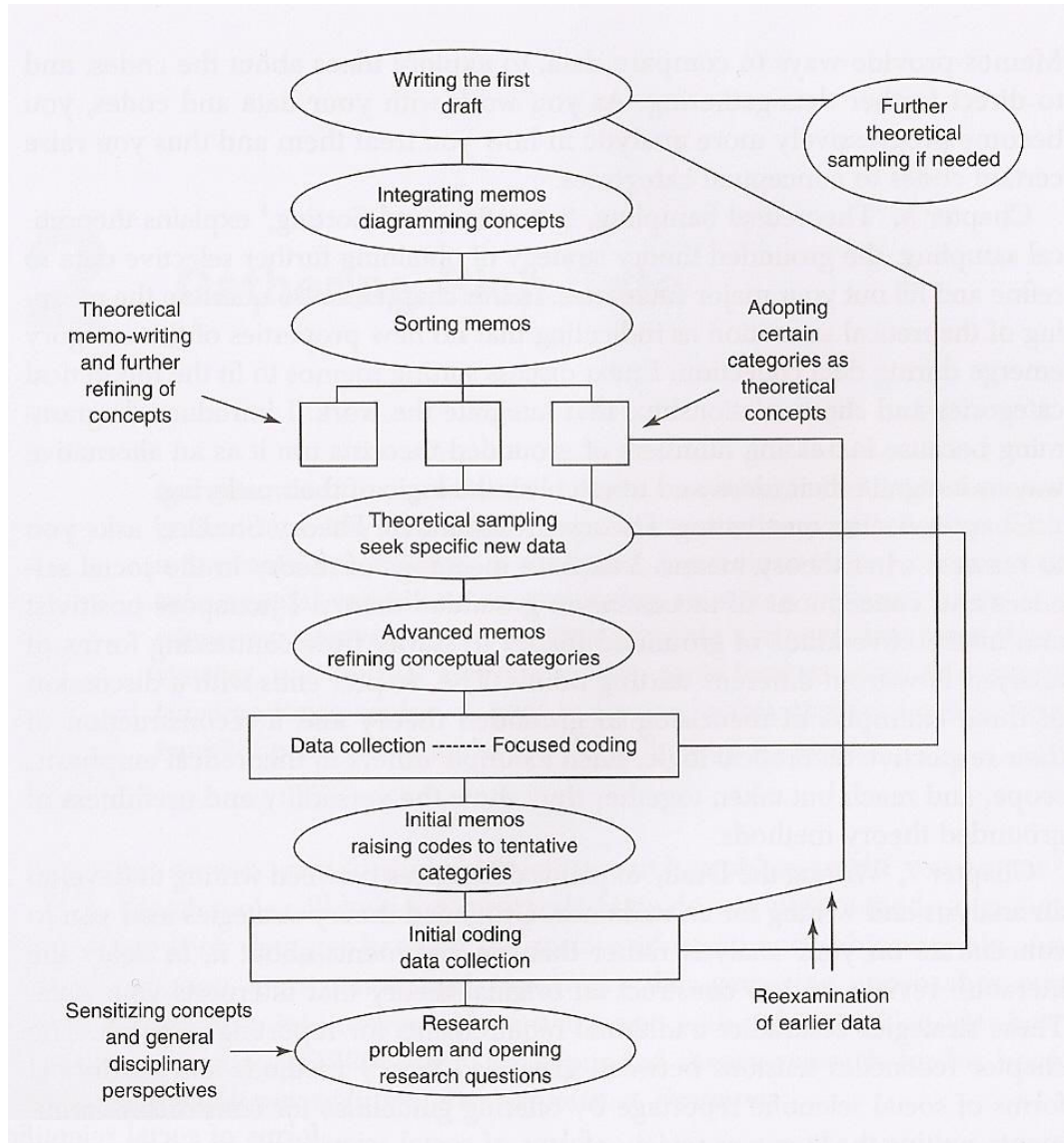
Pt. N°	NRS PH	NRS PT	Sex	Age	Civil status	Diagnosis	OHS Int.	C. Int	NYHA
1	75	20	M	56	Single	Pulmonary Stenosis	2	1	3
2	25	50	M	22	Single	VSD + AI	0	0	1
3	100	80	F	43	Married	Tricuspid Atresia	2	0	4
4	90	50	F	33	Single	Aortic Valve Stenosis	1	0	3
5	50	60	F	20	Single	Tetralogy of Fallot	2	1	2
6	50	30	M	55	Single	Aortic Valve Stenosis	1	0	2
7	100	95	M	21	Single	Aortic Valve Stenosis	0	0	3
8	40	20	M	20	Single	Transposition of great arteries + ASD	2	2	2
9	50	50	M	25	Single	Tetralogy of Fallot	1	3	2
10	70	50	M	35	Single	VSD + Double outlet right ventricle	1	1	2
11	100	0	F	56	Married	Tetralogy of Fallot	2	4	3

OHS: open heart surgery, C: catheterisation intervention, NYHA; New York Heart Association functional class

As can be seen in figure 6.1, there are 4 main stages of analysis in Grounded Theory, which is supported throughout with the writing and integration of memos, which are notes on the research process which outline the analytic process and the map of the path taken. Memos are of two types; initial memos which are short and descriptive and advanced memos, utilised in the discovery of the categories and their evolution and redefinition.

In the process of initial coding, line-by-line coding was done. After that, focused coding was performed by grouping the initial codes, synthesizing and explaining larger segments of the data. Finally the relationship of the categories developed were explored in the process of theoretical coding, leading up to the proposal of a theory. The process was not linear and many times the raw data was referred to as the analysis and interviewing proceeded. Interesting areas which emerged during interviews were explored in the following ones if not brought up spontaneously by the patients (K. Charmaz, 2006). A visual representation of the Grounded Theory process can be seen in the figure below (K. Charmaz, 2006).

Figure 6.1. The Grounded Theory process. Charmaz (2006) pg. 11



6.4 Results

Here are the categories which emerged followed an in-depth description of each one:

Table 6.2. Summary of categories emerged from data

<u>Core Category: Having condition and being different as a consequence:</u> Feeling different and being perceived as different		
<p><u>Outcomes and limitations of having a CHD</u></p> <ul style="list-style-type: none"> • Dealing with physical limits in childhood and adolescence • Sexual and reproductive issues; sexual performance, pregnancy and child rearing • The surgical scar • Rejection, humiliation isolation and perceiving oneself as a burden • Positive outcomes: being pampered and feeling more mature 	<p><u>Memories during hospitalisation</u></p> <ul style="list-style-type: none"> • Difficult moments during hospitalisation and illness: experience of intense suffering • The relationship with sanitary personnel <ul style="list-style-type: none"> ○ Ambivalent feelings of patient towards sanitary personnel ○ Positive handling of the patient <ul style="list-style-type: none"> ▪ Helpful practice during child hospitalisation ▪ Highlighting progress ▪ Treating patients with dignity ○ Negative handling of the patient <ul style="list-style-type: none"> ▪ “Colluding” with the patients’ (implicit) requests: <ul style="list-style-type: none"> ▪ Insensitivity of the sanitary personnel ▪ Misdiagnosis and its catastrophic consequences 	<p><u>Coping strategies</u></p> <ul style="list-style-type: none"> • “Getting used to the inevitable” • Not being alone with this condition • Comparing oneself with others in worse situations • Utilisation of irony and humour • Getting support from family partners and friends • Minimising danger of situation to avoid pain/avoiding unpleasant situations • Blocking out threatening information • Hoping thing will go away on its own • Perceiving oneself as “Being ill” • Defying limits and its possible consequences

6.4.1 Outcomes and limitations of CHD

Having a CHD involves becoming aware of having restrictions others around you do not have, and the process is different for all patients and a strong link emerged between the awareness process and the coping strategies adopted by the patients later on in life. As outlined in the core category, the consequences of having a CHD are not only negative; they also help the patients to have a greater sensibility and maturity and to appreciate life more. It is important to focus on this when providing support to these patients.

A. Dealing with physical limits in childhood and adolescence

The patients' parents and their family play a very important role, in how the patients see themselves, how they feel about themselves and how they behave towards the illness. When the parents of children with CHD do not adequately perceive and communicate the condition to their children this can have catastrophic consequences.

Many patients remember that their parents told them that they cannot strain themselves a lot, and this is when they started to understand that they were different. However in the case of the first patient interviewed (1), when he was a child, since his brother was handicapped and her mother also had a disability, no one ever had the time to take care of him, resulting in him doing manual work, and becoming aware of his condition much later in life when he was 50 years old, even though he felt that something was wrong when he was about 14, he ignored his physical symptoms.

Even because...In a way I couldn't trust anyone...I used to ask my mother and she would say "It's nothing, it's like that...Don't worry it's nothing, it's because you're growing up"

Of course, even my brothers...it's not as if...We were five but not even one of them told me let's see...One didn't care, the other one didn't care and so...(1)

Him actually getting medical help, coincided with finding a partner and his brother passing away; it's as if he was finally able to take care of himself in that moment.

When it comes to another patient (2) he refers to the fact that his father was sporty and that he never told him what he could and could not do. In fact this resulted in him doing what he wanted, and actively looking for medical professionals who supported his view of himself, discarding those who would have told him what he didn't want to hear, as will be outlined in more detail in the following categories.

Some of the patients report that their parents were overprotective and it seems that this led them to do the opposite of what they were told. This attitude was kept also in the future by one patient, confirming the hypothesis that certain upbringing patterns can have an influence on how the patients deal with their illness later on.

my mother used to tell me, don't run, don't play ball, I couldn't do it she always told me but I used to do it...even to work, I used to do tiring jobs...(9)

It is possible that the children's behaviour also influence the parents because this patient in particular says that he was always very lively; so overprotection and being lively become a vicious circle.

It seems that being told that one has a condition when they are very young, in a simple manner, helps them to develop a good awareness, and as a consequence it seems that they can deal with the situation better, as explicated by another patient:

Honestly for me even in hospitals, it was normal, so I can't tell you when I started, that is my mother has always been the type to always say, obviously she wouldn't go in detail, but she told me, you are not a boy like the others, you have a little problem with your heart so you cannot do everything, and as a consequence I don't know if I asked questions, which I probably did, but I think I reacted in this way, remaining tranquil. (7)

In this case, we can see the diametrically opposite position from the first patient; in that case the (1) patient's mother, not only never informed the patient that he had a cardiac condition, but she completely ignored the fact that her son was ill, not checking what was wrong, and this caused many problems to the patient.

In the second case, we can see how the fact that the (7) patient was told that he was different, and eventually he got used to his condition, resulting in a better handling of the condition and with him being more calm, as will be explained more

extensively in the category “coping strategies”. It also useful not forbidding the child to do anything, but suggesting them to do things with moderation, when it is possible.

Having to deal with physical limitations is also present in adolescence when the desire to do what is forbidden and to conform to peer pressure are even more present. In fact some of the patients have a very hard time accepting their limitations during this period. When things are explained clearly and without too much anxiety the patients grow up with an awareness of being different, and being restricted, but it does not make them feel terribly bad.

However, in one particular case (7), after the patient adjusting to his condition by doing very limited physical activities, he had an incident from which they found out that his condition had gotten worse, and that therefore he had to be further restricted. This led him to feel very demoralised and also ashamed because his mother had to carry his satchel to school.

Due the physical limitations the patients sometimes feel they have no choice but not to participate in the activities their peers are involved in and this makes them feel excluded from others. This can result in isolation and choosing to remain at home.

B. Sexual and reproductive issues; sexual performance, pregnancy and child rearing

Most of the men in the sample commented on how having the condition also affected their sexual performance. The implantation of mechanical cardiac devices can give rise to embarrassing situations during intimacy

I noticed that when meeting new people, suddenly my problem came into my mind, that is that with the valve, when one is particularly excited the person close to you can perceive it, so my problem was that capacity the other had to understand...when I used to like someone it used to stop me, that is I had some hesitation and I used to tell myself, how do I deal with this thing? (6)

One patient (7) asserted that he knew that he was not like the others when it came to sex, but he stated that for him it was not a problem and that if his partner loved him she should do so for both his strong points and his week points. He was aware though, that this was a weak point, that he could not “perform like others”.

Another 35-year-old patient (10) describes how he never had sex because both himself and others were fearful because of his condition and that he had never discussed the issue directly with a doctor. This indicates that it is necessary for the sanitary personnel to be more active in informing their patients about their condition and to also initiate discussions which can result as being embarrassing for them.

Whereas sexual performance emerged during the interviews with the male patients, the issue of pregnancy and child rearing was more prominent for the female patients. The only cyanotic patient in the sample (3) always wondered if she could have ever had children and for a while she thought she could. In the end she was advised not to because she could have circulation problems and so she gave up the idea. She tried compensating this by taking care of her friends' babies, and this helped, however when the kids grew up she felt empty. The fact that she could not give a child to her husband also made her feel bad, and different from her healthy friends. The patient also thought about adopting a child but in the end she also gives up on this idea because she believes that legally she would not be a good candidate because of her condition, and she also feels she might be an inadequate mother because of it.

C. The surgical scar

Some interesting gender differences appear when it comes to the surgical scar. The surgical scar is perhaps the most visible sign of having the condition, after cyanosis. The female patients seem to be particularly sensitive about it, and even though through time it seems they bother less about it, a small influence remains; for example in the choice of what clothes to wear. Most of them are bothered when people ask them what happened to them so they wear clothes which cover it up, or they use foundation.

“And then the scar, I didn't want others to see me wearing the bathing costume with the scar from the 12-year-old boys, you start growing up, it was a trauma...I used to see my girlfriends all healthy and myself with a scar from here to here...I used to put on one piece costumes...”

(Later in life)

The sweaters always high necked...but I started wearing bikinis.” (3)

The scar is not only a problem for esthetical reasons in women, it is also the means by which other people notice what the patient could have passed through, and for this reason it is often covered, as reported by 2 female patients (4,5).

When it comes to the male patients, and how they perceive the scar, it seems that they are also bothered by it in the beginning but not as much as the females, also because sometimes there is the possibility to cover it with body hair. Also it seems that in the end one of the patients (6) also got fond of it and saw it as being his “quirk”

Another patient (7) also saw the scar as being part of him, as being part of his story and of his identity and he even grew fond of it and he got upset when his father suggested that it should be removed with plastic surgery.

D. Rejection, humiliation isolation and perceiving oneself as a burden

The physical limitations have often made the patients feel distance between themselves and others and sometimes the others willingly or unwillingly underline this difference. Some of the patients are reluctant to immediately tell their romantic partners that they have this condition, and some patients actually tell of being left because of this condition, either because their partners were in some way scared or because they could not handle it.

Sometimes cyanosis can be the cause of other people picking on the patients. When other people do not understand what the condition is sometimes they can behave in a certain way which cause shame, embarrassment and trauma. In particular, the cyanotic patient in the sample describes two incidents at the sea which depict this.

Once I remember that I was at the sea and there was a group of kids and I still hadn't done the operation, I was always cyanotic and I got to know some kids, they were 12, 13 years old and this kids, we used to play together, once during the evening my cousins went there and they saw a drawing which was me a blue girl with blue glasses, blue, blue...I remember this had hurt me”

Once a woman who was with a girl, who was about 20 years old, held me from my arm while I was going down from the ladder (to the sea) and she said “Come up look at what a violet mouth you have, if you feel bad it will be my problem!....and all the people were looking, for me it was a trauma...(3)

Another feeling the patients had in common was the fact that they were a burden, and that because of their condition they encumbered their family, their parents and their partners with heavy loads. Perhaps because of this, they often feel that they shouldn't share their worries with their parents to make them worry, keeping everything inside. Some of them also refrained from engaging in group activities or sports because they feared how others would feel if they felt bad in their company.

When the patients have a difficult physical situation, and they need to be taken around they can feel that they are a burden for others. The fact that others have to take care of them can make them feel guilty, even though they have no control over their physical health.

E. Positive outcomes: feeling more mature and being pampered

Having the condition does not only entail having limitations but also positive outcomes. Many patients declare that as a consequence of their condition, they feel they have matured more quickly and they can appreciate the important things in life better.

That is in order to deal with these things you have to strengthen yourself, think positively and on the one hand a bit more of maturations, maybe more quickly than others and trusting others, doctors, parents, so for sure the positive things were also more present. (7)

As specified earlier, one patient (3) felt that her husband loved her because she was different, in a positive way, more sensible and reliable. Patients also describe being pampered and given gifts because they had this condition, and some of them (7) also associated going to hospital with being something pleasant because their parents gave them gifts.

6.4.2 Hospitalisation experiences

A. Difficult moments during hospitalisation: experience of intense suffering

Strong adjectives such as “nightmare” and “Calvary” are used to describe the patients’ experiences during hospitalisation. It is not surprising that some of them also describe developing a phobia for white coats when they were children

The whole experience of being hospitalised when the patient was young is described as being a nightmare twice, because similarly to a nightmare things in the memory are fuzzy and she doesn’t know what is going to happen. She cannot make any sense of what is going on, and she is scared about what is going to happen and because she is separated from her mother.

So in the beginning I used to say the nightmare is going to end, that is I have always prayed, I always hoped everything would be OK, even though in the last week before the intervention I was worried, I thought I wouldn’t make it. I was very scared, very
(5)

The word “nightmare” is used also by another patient to indicate the time when she was affected by heavy physical symptoms because of a misdiagnosis, and she was feeling bad but the cardiac surgeon continued to reassure her that she was fine.

Most of the patients feel that they do need support during the whole hospitalisation process and many have emphasises that they need to be supported especially when they are in the intensive therapy unit. They would like to have the company of someone when they come into their senses from after the operation

Referring to the hospitalisation process: *“According to me something which should accompany the whole process, a preparation that is, someone who talk to you before and also the psychological state when you wake up there, there is nobody because there is nobody in the ICU because nobody can be close to you according to me this would be useful”* (4)

More than one patient state that a figure like a psychologist could help them prepare better to undergo the intervention, and could also be an important support for the fear they feel.

One of the things that makes patients feel most uncomfortable during hospitalization is the fact that they are heavily dependent on the nurses for the most basic needs. Because of this they might even not call them because they felt awkward and didn't want to disturb too much.

Once again the theme of the difference with others emerges also during hospitalization; the comparison with other patients who are less dependent than them, can cause the patients to feel worse once they are brought back to the ward from the ITU.

Some patients describe the unpleasant experience of finding out bad news, which change everything. Expressions such as "big blow" "everything went topsy-turvy" (5, 6) describe how the patients feel after the equilibrium they reach cannot be maintained because of the events which occur.

B. The relationship with sanitary personnel: the patients were ambivalent towards the sanitary personnel and they gave examples of positive and negative experiences they had:

- Ambivalent feelings of patient towards sanitary personnel: On the one hand the patients know that their life depends on the sanitary personnel, they are grateful when they get help, on the other hand, the frustration of being unhealthy, needy and dependent may be vented out on the personnel, in the form of: "you are incompetent, you haven't done enough, it's not right etc." Or perhaps, there are certain things which have not been done well and this also exacerbates this feeling, or justifies the venting out which becomes accentuated. The ambivalence towards hospitalisation can be seen in this narrative;

I was treated very well at that hospital for goodness sake, they made my fever go, they told me I had to be operated again so it was fine with me, but when they told me that I had to be operated upon again I said I won't go there, certainly not because if I have to undergo another operation after 4 years when it had to last 15, I won't go (4)

- Positive handling of the patient

1. Helpful practice during child hospitalisation

Interviewing adults on their life experiences connected with the condition of CHD is very useful because they have elaborated their memories, and they can reflect what was good for them, and what helped them, and what didn't, especially when coming to being hospitalised and being operated when they were children.

As described before, the hospitalisation for children can be considered as being a nightmare. One of the patients (3) described how the behaviour of the doctor had put her at ease when she was a child. This entailed:

- Explaining in a very simple way what is going to happen and using diagrams in explaining to the child what is going to be done to them *“Yes there are different ways of saying it... You know, now I have grown older... When I was a little girl the doctor used to tell me, he used to do a little diagram and he used to tell me “Look I am going to perform this operation on you”*
- Showing the child the clinic and the IT unit *“and he used to make me feel less afraid, when I used to go to the clinic he used to show me the ITU, “After the operation you will find yourself here...”*
- Showing the child the operation theatre *“He used to show us the operating theatre and he talked to us about how the anaesthetist came here, he spoke about the hospitalisation, that is according to me this is good but I don't know how they speak to children, according to me it would be useful to talk to them and to show them the ITU”*

Of course the subjective differences in the children's temperaments and personalities need to be taken into consideration, and a tailor made preparation plan should be created for each child, however these could be useful indications for preparing children to undergo operations.

2. Highlighting progress and treating patients with dignity

It is important that the hospital staff is sensitive to certain aspects of the patients, and highlights positive things, and omits to say certain things or says certain things in a very delicate matter, especially during hospitalisation when the patient

needs to be operated upon. For example the cyanotic patient outlines how pleased she was that her lips were pink, and that the nurses kept mentioning it.

People with CHD want to feel that they are normal people, who have to face difficult conditions. They do not want to be identified with the disease, but want to be perceived as being normal people, who have to deal with difficult situations and health problems. When this attitude is adopted by the sanitary personnel they feel better.

- Negative handling of the patient

1. “Colluding” with the patients’ (implicit) requests:

One young male patient exhibited very clearly that he couldn’t accept himself as being, or as having a physical condition. He wanted to do sports at all costs, and he knew that his cardiologist would not have given him permission to do so. So instead of going to him, he went to a GP, “more of an informal chat”, and the GP confirmed that he could do sports, without even doing the necessary check-ups.

Professionals who work in this field should have the capacity to recognise to prevent things such as this from happening and help the patient to elaborate a more accurate sense of self, trying to find an equilibrium between not demoralising the patient too much, but at the same time, making them aware of the possible risks that some inadequate behaviours (such as physical activity) can have. When I asked him why he did not go to the cardiologist instead of the GP his reply was:

because the cardiologist would have told me that I couldn’t do anything. Like the cardiologists of the gym X, they told me I couldn’t play football, because if you then have problems...the cardiologist would have told me the same thing for sure. (2)

Sometimes it appears that the patients do not want to know what the operation entails and what the consequences will and can be; this could part a defence mechanism which allows them to handle the situation. It would be better to support these patients, and let them know, in a delicate manner what is going to happen, otherwise they will probably suffer the consequences when they awaken after the operation, as a patient (6) clearly describes.

At first it seemed that the patient didn't want to know much about the intervention, because he could not handle it, and this resulted in him feeling very bad when he woke up. The sanitary personnel should be aware that patients could not want to know details about the operation because they are scared, and that this could have negative consequences when the patients recover from the operation.

after the intervention as I was waking up I asked myself, you didn't want to know what will happen to you, how your life will be, how your future will be, that is what this operation will entail, what you will be able to...and in that moment I regretted not being updated on what had happened to me. (6)

A psychological preparation from the beginning of the hospitalization could help prevent this from happening.

2. Insensitivity of the sanitary personnel

If the medical staff is insensitive, this can cause a lot of problems and a lot of fear in the patients, especially in very delicate moments such as just before a procedure, especially if that procedure has not been done before by the patient, and there is a lot of fear. One patient describes being told about how risky the operation is in a very insensitive way by a nurse:

well one person passed away whilst he was doing it, he was 25 years old. I did not speak. "but the operation is hard and many don't make it" (3)

From this example the necessity of the sanitary personnel receiving training in the psychosocial handling of the patients is quite evident.

Other examples of sanitary personnel being crude to the patients also emerged from the interviews with other patients. For example, in one occasion, a person who had just been operated, was told by trainee doctors that she just need to be operated again, very crudely, and the patient felt as if she "wanted to die". Not only that, her parents obviously also felt very bad as a consequence.

“no you have to be re-opened”, and he said it exactly like that, I mean right after I underwent an intervention of that kind...I wanted to die” (4)

In fact the patient reports that she felt as if she wanted to die she felt so bad.

One of the patients (6) decided to be followed by the doctor who operated him and who had specialised abroad, even though he lived far away from him. It seems that this doctor had some problems when he came to be reinserted in Italy because of the different procedures he used to use, and when the patient went to do some tests in other hospitals, he used to get treated badly when the doctors knew by whom he was followed; he describes how the doctors did not approve of his therapeutic choices and that they were aggressive and offensive.

3. Misdiagnosis and its catastrophic consequences

The CHD population is a very particular population, and it is necessary that specialists who are capable to diagnose conditions correctly work with them, otherwise there can be catastrophic consequences as described by one patient (5), who even though was manifesting many physical problems, was told that “she had the heart of a lion”. It is interesting that even though this had extremely negative consequences on her life, she is very controlled when expressing her anger.

The same patient describes how the cardiac surgeon who had misdiagnosed her tried to hide his incompetence by blaming the family for neglecting her.

The incorrect observations about the mechanical valve of one of the patients (6) by some doctors caused could have cause the patient a lot of worry, and it caused him to get very angry when he realised that they were not competent to make these comments; he was told that his valve had a wing which didn't function well to find out later that his valve only had one wing.

This further highlights the necessity for the following:

1. Cardiac surgeon should have the necessary competences to treat the CHD population
2. If not, they should assume the responsibility to acknowledge that they cannot take care of the patient, and they should be able to be able to refer the patients adequately.

6.4.3 Coping strategies

A. Accepting the inevitable

One of the patients provides a very touching description of this phenomenon in the beginning of the interview. He describes how he has always felt different and limited, and that this has caused him suffering, but at the end of the day, one accepts the inevitable and moves on.

since always, I was born this way, so since I have never led a normal life, as a healthy boy, I never thought about the possibility “if I hadn’t have got sick I wouldn’t have this problem”, I always though, I have it if I live with it it’s better for me, I accept the inevitable.” (7)

Even when things got worse for this patient, the fact of accepting the inevitable seems to make suffering more bearable.

It could be possible that since some patients are born with this condition, and they are used to having it since when they are born, they perceive certain limitations as not being so hampering; many patients describe doing their activities and sports and stopping when they feel tired, even though this could be an attempt to downplay their situation in an attempt not to feel so different.

B. Not being alone with this condition and comparing oneself with others in worse situations

The fact that there are other people who have suffered the same thing, who have the same kind of scar, makes these patients feel better. In fact when I used to go up in the ward and I used to talk about the patients many of them said that it was a relief for them seeing that they were not the only ones, and sometimes seeing people who were in a situations which was worse than theirs helped them appreciate that after all not everything was bad.

(on getting over having a scar) in that period I got to know another girl who had also had a heart operations and she had a scar like me and this made me think, ah I'm not the only one (3)

Saying to oneself that things could be worse and that there are other people who have worse conditions seems to be a good strategy to mitigate negative emotional states. Sometimes not only imagining that there are people out there with worse conditions, but also having firsthand experience of people who they perceived as being worse, and who eventually passed away was a source of inspiration and strength.

I always told myself that there are worse cases in life than mine because I had an ugly bereavement in my family and the courage of the person who I lost used to give me strength (5)

Another patient (6) compares himself with other patients who were brought in by the ambulance, whereas he went there with his own two feet, and he could decide when to do the intervention.

In the end he states that he believes that there are others who have suffered much more than him, because he was affected by his condition much later in his life, and he saw patients who were 14 and 18 years old in the ward, which almost made him feel guilty for the way he felt.

C. Utilisation of irony and humour

The utilisation of irony and humour can be a great resource to deal with discomfort caused by the condition. One of the patients handled feeling different from others because of the “tic” of the mechanical valve by using irony; a strategy which seemed to allow him to handle the situation with much more ease.

by using irony, that is I didn't use to say anything and then depending on the person, their capacity and cultural level, I used to invent stories (patient giggles) even which were very imaginative, I also told a very simple girl that I was an alien (patient giggles again). (6)

D. Getting support from family partners and friends

The support of family and partners seems to be essential for these patients; in fact as already outlined previously, the lack of this support led one patient (1) to completely deny his condition and not take care of himself. It is not a coincidence that he found the strength to face reality when he also got to know his partner.

Nearly all patients outline how important their parents, partners, relatives and friends have been, even though sometimes the parents can provide extra stress during hospitalisation because they do not manage to deal with their own anxiety. For this reason it is also important to supply psychological support also to the parents and relatives visiting the patients, in order to help to be a resource for the patient as much as possible.

E. Downplaying the condition: from minimisation to complete denial

Often in these patients we can see that they minimise their physical limitations, downplaying how serious they are, or their effects on their life. This phenomenon can happen at different levels of awareness and a patient (9) described it very accurately by saying that he used to feel “ill” in inverted commas.

Sometimes the patients cannot handle knowing what the surgical operations they have to undergo entail, and what consequences they could have, and they chose not to ask about them or not to let the sanitary personnel explain things to them clearly. This can leave the patient with a sense of anxiety once they recover from the operation as already outlined previously

Many times the physical symptoms are ignored, until something quite serious which cannot be ignored happens. The mistaken belief that things will get better on their own is a false belief held by quite a few of the patients, and their relatives, especially at the beginning of the illness, possible because it is hard for them to face such a condition, but this erroneous perception could also have been caused by a misdiagnosis.

For example one patient (6) who knew about his condition, waited all night before checking out what the problem with his breathing was. It appears to be quite strange that he didn't attribute this to his heart. In another case, a patient (1) used to

feel that something was wrong, but he used to ignore his physical symptoms, and continue doing manual jobs. This has even lead (together with other factors, such as his mother and siblings ignoring it and a low level of education) to the discovery of the condition much after in his 50s with consequent catastrophic consequences on his health. Yet another patient (11) did not connect physical symptoms to her cardiac condition, when they were actually caused by it and they eventually led to her hospitalisation.

Also as outlined previously, one young male patient (2) had clear indications from his cardiologist and some sports institutions he could not do any sports but he ignored them, because he wanted to do sports at all costs. He was aware of these limitations, and because he needed some reassurance, he went to a GP, who told him he could do sports, without visiting him adequately, avoiding to go to a cardiologist, so as not to hear what he didn't want to hear; that at that point in time he wasn't fit enough to do sports

F. Defying limits and its possible consequences

Many patients describe this coping strategy; testing the limits, seeing until what point they can do what they are not supposed to do in order to prove to themselves that they are not really that ill, and that different from others. This was described very accurately by one of the patient:.

And I used to tell myself, the illness, that is I have to win the illness anyway, the illness cannot impose on me what to do if I don't want to do it and so I'll do some things and I'll see until what point I will arrive, this was my reasoning.

On the one hand I tried to combat the illness doing the contrary of what I had to do and on the other hand I corrected myself alone. (4)

Also another patient (10) describes this phenomenon with great clarity, connecting this to ignoring physical symptoms:

I used to abuse my limits as always, I have always been an idiot, I have always abused my limits and I always ended up here, and the third time that I did the operation I didn't abuse my limits any more. (10)

This can be considered as going one step ahead from perceiving oneself as not being really ill; engaging in behaviours to prove to themselves that they are not really ill and that the cardiac condition does not really limit them, even though most of the time we can see an awareness that what they are doing is not exactly right. This battle, this defiance of the limits may be more prone during the adolescent and early adulthood phase, after it seems that the patients feel a bit resigned and adjust to their condition, also because they often have scary experiences.

A patient (10) explained very clearly that he used to enact these behaviours because he was feeling well and he didn't want to feel he was an invalid person. This patient seems to describe another face, a consequence of the defiance of limits; that you stop only once you reach the "depths of despair", that you truly interiorise that you are different only when life puts you in front of undeniable evidence, that forces you to readjust

Of course it would be more functional that the patients become aware of this mechanism before they touch the pits of despair or the bottom. "Touch the bottom". The defiance of limits is also clearly shown in the last patient interviewed (11); not only she specified that she used to do behaviours which were forbidden to her, but that she did not take into consideration anything the doctors told her and did the exact opposite; she had manual tiring jobs, and two children.

6.4.4 Core category: Being different

Being different from others who are healthy and do not have the condition is the core category which emerges strongly in all interviews with the patients; it is the core category because it lies at the basis of the main three categories. The consequences (which are not necessarily negative) and limitations due to their condition reminds the patients that they are different from other people, and so do their memories of all their hospitalization; the coping strategies adopted are a reaction to this perception and the associated feelings related to it.

Sometimes the patients also refer specifically to feeling different, as they compare themselves to others. For example, one patient (4) used to compare herself to other children when she was a child, and that she perceived a difference between herself and the others, because she could not do physical activities, and also because she had to go to hospital for checkups, and the other children didn't have to.

However, when she is asked directly if she felt different from the others during this time period (5-18), she says that this didn't make her feel substantially different from others, which could possibly be an attempt to minimise her condition, and feel like the others. Another hypothesis could be, that having these check-ups done so frequently at such a young age, makes the person get used to them, and thus they are seen as not being anything special.

The most clear and touching quotation of being different was provided by a 21-year-old male patient, who had quite a serious condition. Once the physical limitations of a patient increased so much that he felt a greater difference between himself and the others, and he wondered what he was going to do.

Whereas before I wasn't that bothered that I had heart problems, this thing over here differentiated me completely from others and I started to feel a bit sad, and so I thought "what shall I do now? Am I going to remain like this all my life? (7)

In fact this difference for him was too heavy to bear, and he was ready to risk everything in order to gain at least a minimum level of physical strength which would have enabled him to do the basic things he was used to doing, even undergoing a heart transplant.

Feeling different and being perceived as being different are obviously connected, because the construction of our self-perception greatly depends on the feedback of others. Patients often report the others seeing them as being different, but not only in a negative way. An interesting thing about CHD is that the illness is not usually apparent to others, unless there is a visible scar, or the patient is cyanotic.

The only cyanotic patient in our sample (3) specifies that it was more the others' perceptions of herself than her own perceptions which made her feel different. In fact she says that she couldn't see her violet lips unless she looked at herself in the mirror, but others pinpointed these differences on several occasions. The same patient

reports of how her partner, who later became her husband, perceived her as being different in a positive way, as being more reliable than the other girls.

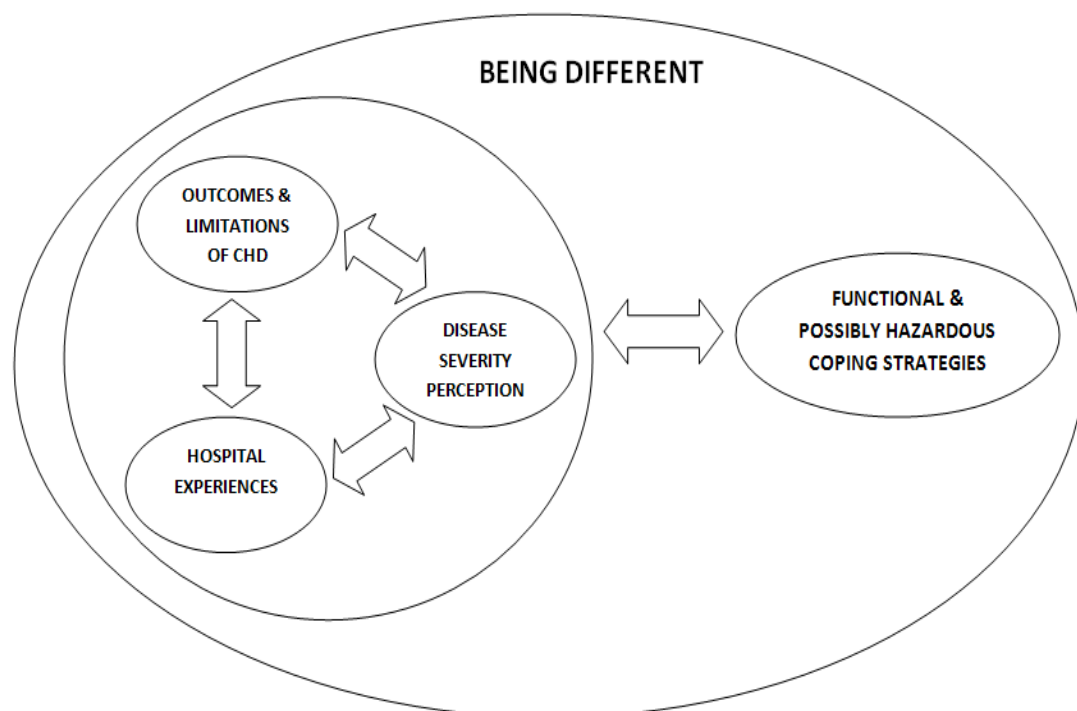
The patients are often concerned about how others will view them, especially because CHD is not a condition which is well known. In fact some of them show concern at the possibility of being seen as crippled, and some others feel angry because the ignorance of other people caused embarrassing situations which made them suffer a lot.

The different social settings of the patients also bear an important influence on how they feel. One patient (10) lives in a small town and he describes very clearly how all the inhabitants see him as being different after he felt bad, and he had to be carried to hospital by a helicopter; for everyone in town he is the boy who “lives between life and death”.

6.5 Discussion

The categories that emerged and the relationship between them can be viewed in this diagram:

Figure 6.2. Visual representation of Grounded Theory categories



Also in this research, the fact that some patients do not have a realistic perception of the severity of their condition emerges clearly. The analysis of these patients experiences can give us some hypothesis regarding of why this occurs; as confirmed by the literature reported in the previous chapter, the fact of being different emerges strongly and this underestimation could be an attempt to “normalise” the situation as explained by Claessens (Claessens et al., 2005).

In this study an interesting comparison could be done between the quantitative data and the qualitative data. The patients who gave a similar score to the physician actually showed an increased awareness of the severity of their condition but not only that, they seemed to utilise coping strategies which were more adequate.

It was very interesting to see the clear connection between how the upbringing of the patients, their social status, education and the context where they were brought up had a deep impact on the coping strategies utilised. The ones which emerged from the data can be categorised and assessed according to the classification provided by Moos outlined in the previous chapter (Moos & Holahan, 2007).

Table 6.3. Application of coping strategy classifications to sample data

	Approach	Avoidance
Cognitive	<p>Cognitive approach coping</p> <p>Not being alone with this condition and comparing oneself with others in worse situations</p>	<p>Cognitive avoidance coping</p> <ul style="list-style-type: none"> • “Accepting the inevitable” • Utilisation of irony and humour • Downplaying the condition: from minimisation to complete denial
Behavioural	<p>Behavioural approach</p> <p>Getting support from family partners and friends</p>	<p>Behavioural avoidance coping</p> <p>Defying limits and its possible consequences</p>

Confirming the literature, the patients themselves describe avoiding coping strategies they had utilised previously as not being adaptive, especially when it comes to the defiance of limits. Other avoidance strategies, such as accepting the inevitable and the utilisation of humour did seem to be adaptive from what they 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 lead the patients not to acknowledge cardiac related symptoms which could have put their health in danger.

It is interesting to note that the patients (1, 11) who underestimated their conditions the most as shown by the comparison between the NRS scores 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 utilising mostly cognitive and behavioural avoidance coping strategies.

6.6 Methodological issues

As explained previously ACHD patients are a very heterogeneous group. Although a range of severities and conditions was included in the sample, only one patient with cyanosis was interviewed, however saturation was achieved after interviewing 11 patients. Although the results of the present study cannot be generalised to the entire populations they do give useful indications on the psychosocial handling of these patients, especially when it comes to their hospitalisation.

6.7 Conclusions

Once again, the correct perception of the severity of an illness was paramount in handling having a CHD, an underestimation of the condition is associated with maladaptive coping strategies which could lead to risky health behaviours. It is essential to help these patients to get a more accurate perception of their condition and to try to discover, together with them adaptive coping strategies to increase their

compliance with the medical team and to help them deal with their condition more effectively, also by empowering them to use the resources which are already available to them.

Chapter 7

Discussions and conclusions

7.1 Disease severity rating, HRQoL and life experiences in ACHD patients

The main focus of this dissertation, was to examine how ACHD patients perceive themselves in their illness, in particular the severity of their condition, and the possible effects of a distorted disease severity rating on the patients' psychological well-being during hospitalisation.

The literature has already outlined that there are significant gaps in what the patients know about their condition (Chessa et al., 2005; Moons et al., 2001), however in the studies concerning QoL and psychological functioning, this has only been marginally addressed and no direct comparison between patient severity as rated by the patients and by the medical physician has been done.

Moreover, in this dissertation, these two evaluations were both compared with quantitative and qualitative data, giving rise to some very interesting considerations. First of all, when comparing the two scores, it seems that the patients who have the hardest time to give an accurate perception of their condition are those with medium severity cases, and even though when the patients had a severe or mild condition there was better agreement between the measurements, a tendency of the patient to overestimate the severity of the disease for low values and to underestimate it for higher values was verified.

Furthermore disease severity rating as done by the patients was considered to be the only predictor when it comes to severe distress and also general hypochondriasis; this has very important clinical implications. Psychological interventions are paramount in those occasions when the patients rate themselves as being very severe and who in reality are not, because this discrepancy can cause a lot of distress which can be alleviated.

Although the denial of one's condition can be a functional coping mechanism during certain stages of the illness and it could allow the patients to adjust, an extreme

underestimation of one's severity could have very detrimental effects on one's health as outlined very clearly by the qualitative study, presented in this dissertation.

The last patient interviewed, sustained that her severity was 0, and the physician evaluated her severity as being 100; meaning that there was the maximum discrepancy possible. It was clear that the patient dealt with her condition using mostly avoidance coping strategies, and this has led her to put her health at risk on many occasions. Whereas some of the patients themselves, identified and acknowledged that in the past, especially when they were adolescents, they enacted these kind of coping mechanisms, but then stopped when they became aware of how dangerous they were, this patient in particular, showed no such awareness.

The qualitative research in this dissertation confirmed the findings in the literature; the feeling of being different because of the condition is the main issue when it comes to ACHD patients. This is not only the source of negative consequences; as many patients outline, it has made them stronger, mature more quickly, it has led others to see them in a certain light and chose them as partners and finally it has also led them to be pampered by others.

Obviously the negative consequences also emerged, and they included isolation, fear, being ridiculed, having problems in many social spheres, such as finding an occupation and a partner. Some interesting themes which emerged, which had not yet been dealt with in the literature are;

- **Difficulties in sexual performance, when it comes to the males with CHD:** There has been some research on how women feel about their body image and about pregnancy issues (Claessens et al., 2005; L. T. Gantt, 1992), but no other qualitative research has outlined the difficulties which these male patients said they encountered; in particular the fear of not being able to perform like other men and also the fear of being ridiculed if they have cardiac devices implanted.
- **Particular difficulties during recovery in the intensive therapy unit:** quite a few patients outlined that the most difficult time during hospitalisation was after the operation, when they were intubated and they could not move and they felt very helpless.
- **The link between the underestimation of the condition and maladaptive coping strategies:** the direct comparison between disease severity perception and evaluation, and the utilisation of these scores together with the quantitative data

have outlined that an inadequate perception of one's severity can lead to maladaptive coping strategies which can put one's life at risk.

7.2 Clinical implications for the psychological handling of ACHD patients

In order to improve the patients' rating of severity, the educational "tool kit of information" suggested by the Adult Congenital Heart Association (ACHA) (*Adult Congenital Heart Association*, 2008) which contains suggestions about what every ACHD patient should know about their condition, should be taken into consideration. This refers to the ACHD patients':

- Ability to name and describe defect and interventions
- Ability to recognize cardiac symptoms
- Understanding of need for on-going care
- Information on how to find appropriate care
- Understanding of risks particular to defect(s)
- Understanding of risk of CHD recurrence
- Knowledge of appropriate birth control options
- Understanding of pregnancy risks/ special needs
- Understanding of appropriate exercise activities
- Access to medical records
- Access to appropriate vocational education
- Access to support and on-going information

It is important to work together in a multidisciplinary team to make sure that the patients acquire this knowledge and have access to medical records, updated information and also appropriate vocational training.

As stated previously there are 4 main theoretical and methodological approaches typical of clinical psychology, which can be considered as the basic elements in "Psycho-Cardiology" (Molinari et al., 2006) and these shall be used as a theoretical framework in order to present the implications for the psychological handling of ACHD patients, which emerged from the literature examined and also both quantitative and qualitative empirical findings.

7.2.1 Patient focused therapy

As outlined by the literature and also by both empirical studies, ACHD patients have to overcome a lot of difficulties, and many of them have a very hard time during hospitalisation due to a variety of factors; their health condition, their family and possible problems with the sanitary personnel (Kools et al., 2002; Kovacs et al., 2005). Often the patients have to undergo heavy intervention, make difficult decisions, adjust to abrupt changes in their health and also withstand uncertainty.

It is therefore important to provide support to these patients and this type of therapy supplies the clinician with instruments which help the patient to open up and feel relief. The therapeutic techniques which are typical of this type of therapy would be useful especially when first contacting the patient, in order to create a good working alliance and also when the patient is passing through hard times, therefore before the interventions and during their stay in the intensive therapy unit when they are feeling helpless.

7.2.2 Positive Psychology

One of the major difficulties in CHD patients is their transition from adolescence to adulthood many adolescents who do not manage to successfully transition to adult care (Reid et al., 2004).

The literature also outlines the patients' difficulties with employment and health insurance (Fekkes et al., 2001; Kamphuis, Vogels et al., 2002; van Rijen et al., 2003) which could lead to depression (Popelova et al., 2001).

It is therefore important to keep in mind the resources these patients have and not to focus too much on what is wrong, and to utilise strategies such as empowerment (Kovacs et al., 2006). In a research carried out by Moons and colleagues the most important determinants of an ACHD, QoL resulted as being dependent upon family, job/education, friends, health and leisure time (Moons, Van Deyk, Marquet et al., 2005), therefore it is important to investigate these areas, and potentiate as much as possible the resources already available, in order to increase QoL and to allow the implementation of functional coping strategies.

7.2.3 Cognitive-behavioural therapy

Although a non-directive stance is useful to help the patient open up and to provide support, often in the hospital setting it is necessary to reach certain objectives, such as evaluating a patient and working on therapeutic alliance. Although for some patients encouragement and support are enough, some will need a more direct approach which includes behavioural strategies in order to maximise adherence.

Some cognitive-behavioural techniques are particularly indicated for some problems which this population present, these include increases sensitivity to heart-focused anxiety and bodily sensations (Eifert et al., 2000; Rietveld et al., 2004; Utens, Bieman et al., 1998) communication problems with family, peers and sanitary personnel, getting used to new cardiac devices and also pre-interventional anxiety. The techniques which could be of used are autogonous training, progressive relaxation and social skills training (Kovacs et al., 2006; S. F. Sears, Jr. et al., 1999; S. F. Sears et al., 2004; Seskevich et al., 2004).

7.2.4 Systemic family therapy

From both the literature and the patients' interviews, the fact that some patients' parents can be overprotective has been underlined (Brandhagen et al., 1991; L. T. Gantt, 1992) and this could cause these patients to develop and maintain dependent lifestyles (Kokkonen & Paavilainen, 1992) even though the situation seems to improve as they grow up (Fox, 2002).

The family plays a very important role in the psychological functioning and the QoL of these patients; as outlined by the qualitative research both overprotection and also neglect can lead to the adoption of maladaptive coping strategies. Systemic family therapy can be useful in order to assess how the family adjusts to the illness and it can suggest ways to improve coping and help patients to transition more efficiently into adulthood.

7.3 Limitations

As mentioned previously, when it comes to the quantitative research outlined, the sample needs to be increased in order to make sure that the results are generalised. The physician based her judgement on a set of criteria when giving her evaluation of the severity of the CHD conditions, but these criteria should be further specified in order to increase validity. When it comes to the qualitative research perhaps more patients with cyanosis should have been included in the sample, even though saturation has been reached.

7.4 Future research and final considerations

This research is the beginning of an effort to address the psychological aspects in ACHD patients. Future research topics include the patients' psychological functioning and QoL at follow-up, in-depth research on the transition between adolescence and adulthood and also the impact of this condition on family members. It will also be important to address the difficulties encountered by the sanitary personnel when handling this population.

Some indications have been gained about how to proceed when providing psychological support, but research will also have to be done on psychological interventions specifically for ACHD patients, including also the effectiveness of these interventions.

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