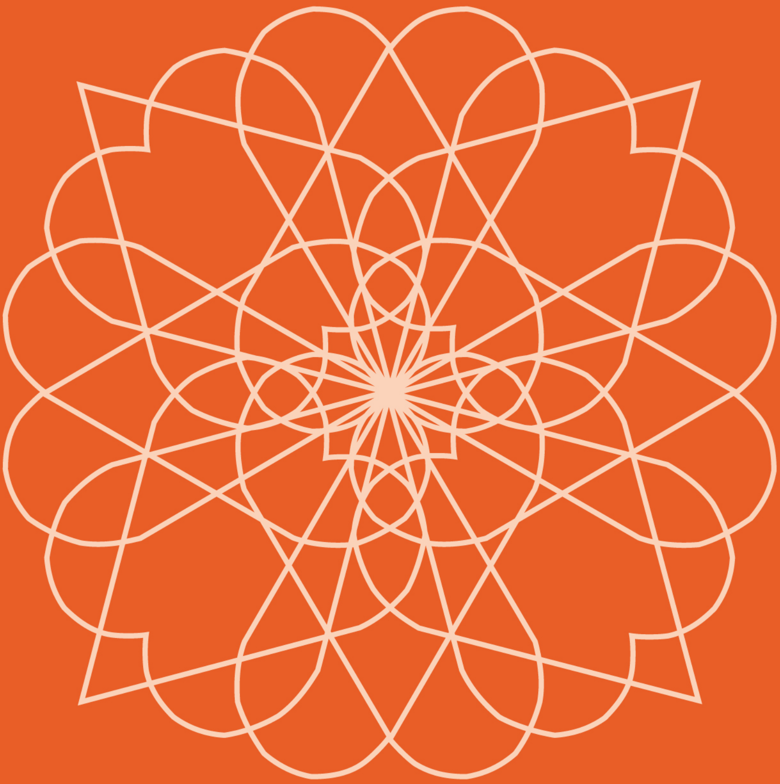
Psychosocial aspects of congenital heart disease in adulthood
A longitudinal cohort study of 20-33 years follow-up



Elisabeth H.M. van Rijen

Psychosocial aspects of congenital heart disease in adulthood

A longitudinal cohort study of 20-33 years follow-up

Elisabeth Helena Maria van Rijen

ISBN 90-6734-319-6

Printed by Optima Grafische Communicatie, Rotterdam, the Netherlands
Figure cover: Susan van Rijen

The study reported in this thesis was performed at the Department of Child and Adolescent Psychiatry, Erasmus MC – Sophia Children's Hospital Rotterdam, the Netherlands.

Rotterdam, 2003

Psychosocial aspects of congenital heart disease in adulthood
A longitudinal cohort study of 20-33 years follow-up

Psychosociale aspecten van een aangeboren hartafwijking op volwassen leeftijd
Een longitudinaal cohort-onderzoek na 20 tot 33 jaar

Proefschrift

ter verkrijging van de graad van doctor aan de
Erasmus Universiteit Rotterdam
op gezag van de
Rector Magnificus

Prof.dr.ir. J.H. van Bemmel

en volgens besluit van het College voor Promoties.

De openbare verdediging zal plaatsvinden op
woensdag 19 november 2003 om 15.45 uur
door

Elisabeth Helena Maria van Rijen
geboren te Gilze-Rijen

Promotiecommissie

Promotor:	Prof.dr. F.C. Verhulst
Overige leden:	Prof.dr. W.A. Helbing Prof.dr. J.R.T.C. Roelandt Prof.dr. J. Passchier
Copromotor:	Dr. E.M.W.J. Utens

Financial support by the Netherlands Heart Foundation for the publication of this thesis is gratefully acknowledged.
The study described in this thesis was supported by a grant of the Netherlands Heart Foundation (grant number 99.033).

Paranimfen:	Cecile van Rijen Paul van Rijen

Contents

1 Introduction

Chapter 1

Introduction

A new patient population

Adults with congenital heart disease form a rather new phenomenon. Improvements in surgical techniques over the last decades have lead to lower mortality rates for children born with a congenital heart disease, enabling more of them to grow into adulthood (Sparacino, 1994; Wren & O'Sullivan, 2001). Today, health care professionals encounter the first generation of adults with congenital heart disease.

Adults with congenital heart disease do not only differ in medical status and history from the well-known cardiac population with acquired cardiac diseases, but also show different psychosocial characteristics and needs. Firstly, the age at onset of the cardiac disease differs considerably for adults with congenital heart disease compared to adults with acquired cardiac disease. For adults with congenital heart disease, the cardiac anomaly may not only affect their present adult life, but it may also have left its mark on their childhood. Experiences of growing up with a congenital heart disease might influence the patient’s functioning at adult age. Secondly, when growing up into adulthood, patients may be confronted with new problems associated with congenital heart disease in adulthood. These problems might concern finding a suitable employment or getting a mortgage (Allen et al., 1992; Celermajer & Deanfield, 1993; Gersony et al., 1993; Hart & Garson, 1993; Hellstedt, 1994; Mahoney et al., 1991; McGrath & Truesdell, 1994) or they may concern the experience of uncertainties or restrictions regarding sexuality and childbearing (Gantt, 1992; Mahoney et al., 1991; Somerville, 1998; Tong & Sparacino, 1994). Adults with congenital heart disease are in the disadvantageous position of being the first generation to experience these problems, since little is yet known about relevant medical and psychosocial aspects. For these reasons, adults with congenital heart disease should be considered as a new and distinct patient population with unique characteristics. Therefore, the medical course of congenital heart disease into adulthood, as well as the psychosocial development of adults with congenital heart disease, deserve special attention.

Congenital heart disease in childhood and adolescence

So far, research into psychosocial consequences of congenital heart disease mainly focussed on children and adolescents (Alden et al., 1998; DeMaso et al., 1990; Gupta et al., 1998;

Kramer et al., 1989; Spurkland et al., 1993; Utens et al., 1993) and their parents (Carey et al., 2002; Davis et al., 1998; Gardner et al., 1996; Lawoko & Soares, 2002; Stinson & McKeever, 1995; Tak & McCubbin, 2002; Utens et al., 2000a; Utens et al., 2002).

Higher levels of emotional and behavioural problems were found for children and adolescents with congenital heart disease in comparison to the normal population (Gupta et al., 1998; Kramer et al., 1989; Utens et al., 1993). Mainly internalising problems were reported, such as medical fears (Gupta et al., 1998), feelings of anxiety (Gupta et al., 1998; Kramer et al., 1989; Utens et al., 1993), depression (Gupta et al., 1998; Utens et al., 1993) and inferiority (Kramer et al., 1989). To a lesser extent, externalising problems in children and adolescents with congenital heart disease were reported (Kramer et al., 1989; Utens et al., 1993). Although in some studies no differences were found in psychosocial functioning between different cardiac diagnostic groups (DeMaso et al., 1990; Utens et al., 1993), several studies showed an association between poor psychosocial functioning and impaired physical functioning (DeMaso et al., 1990; Kramer et al., 1989; Spurkland et al., 1993).

Regarding parents of children with congenital heart disease, psychological distress has frequently been observed (Davis et al., 1998; Gardner et al., 1996; Lawoko & Soares, 2002; Utens et al., 2000a; Utens et al., 2002). In the interaction between parents and children with congenital heart disease overprotection (Carey et al., 2002; Linde et al., 1966) as well as low levels of positive affect and engagement in parents (Gardner et al., 1996) have been reported. Several studies demonstrated an association between parental stress and emotional and behavioural problems in children with congenital heart disease (DeMaso et al., 1991; Gupta et al., 1998; Visconti et al., 2002).

Congenital heart disease in adulthood

Little is known about psychosocial aspects of congenital heart disease in adulthood. Nevertheless, congenital heart disease may play a role in many aspects of the patient's adult life. In this thesis, the following topics will be discussed. A brief outline of what is known from previous studies in presented here.

Biographical characteristics and social functioning

Research into psychosocial functioning of adults with congenital heart disease has yielded ambiguous results. Regarding educational attainments, lower (Kokkonen & Paavilainen, 1992) and similar (Shampaine et al., 1990), as well as higher (Brandhagen et al., 1991; Gersony et al., 1993; Ternestedt et al., 2001) educational levels compared to the normal population were found in adults with congenital heart disease. Although overall favourable outcomes were found for occupational status of adults with congenital heart disease (Brandhagen et al., 1991; Shampaine et al., 1990; Ternestedt et al., 2001; Utens et al., 1994), problems with seeking employment were also reported (Celermajer & Deanfield, 1993; Mahoney et al., 1991; McGrath & Truesdell, 1994). Ternestedt et al. (2001) found normal living conditions for adults with congenital heart disease, whereas Kokkonen & Paavilainen (1992) and Utens et al. (1994) showed that, for specific age ranges, adults with congenital heart disease were living with their parents more often than reference peers. Gersony et al. (1993) found indications for a slight delay in getting married in patients with congenital heart disease. Also, a higher proportion of females with congenital heart disease was found to be childless compared to the normal population (Gersony et al., 1993). Finally, social functioning of adult patients with congenital heart disease as studied by Utens et al. (1994) was considered to be normal, whereas Kokkonen & Paavilainen (1992) found a delay in social maturation. The diverging findings of existing studies into psychosocial functioning of adults with congenital heart disease are due to several factors, such as variation in methods across studies, varying sample sizes with often low response rates, non-standardised assessment procedures and heterogeneous sample composition.

Emotional functioning and psychopathology

As far as emotional functioning is concerned, Brandhagen et al. (1991) reported elevated levels of psychological distress in adults with congenital heart disease. In contrast, Utens et al. (1994) reported more favourable results on neuroticism, hostility and self-esteem for adults with congenital heart disease, compared to a reference group. Besides a favourable emotional outcome, Shampaine et al. (1990) found low levels of adaptability to change, less leadership skills and a high level of harm avoidance. In contrast to the emotional and behavioural problems found in children and adolescents with congenital heart disease (Gupta et al., 1998; Kramer et al., 1989; Utens et al., 1993), the few studies into psychopathology concerning adult patients scarcely reported unfavourable outcome for adults with congenital heart disease (Cox et al., 2002; Utens et al., 1998a). The individual course of psychopathology from childhood into adulthood has not yet been examined longitudinally in patients with congenital heart disease.

Coping styles and social support

Since patients with congenital heart disease may encounter various problems when entering adulthood, it is worthwhile to know what coping strategies they use. Although overcompensation and denial of problems associated with the congenital heart disease have been mentioned earlier (Ternestedt et al., 2001; Utens et al., 1996; Utens et al., 1994), coping styles in adults with congenital heart disease have not yet been structurally explored. In addition to coping styles applied by patients themselves when facing problems, it is important to know how they are supported by their social environment. The role of the social environment has been examined for children with congenital heart disease (DeMaso et al., 1991; Gupta et al., 1998; Visconti et al., 2002). The social environment of adults with congenital heart disease, in which the role of parents is generally diminished, did not receive much attention so far.

Subjective health experiences

The way adult patients feel affected by the congenital heart disease often is not a clear reflection of objective medical measures. As can be expected, in patients with complex congenital heart disease, poor subjective health status concerning issues like physical functioning and vitality has been reported (Greenwood, 2000; Kamphuis et al., 2002). Ternestedt et al. (2001) and Lane et al. (2002), however, both found poor subjective health experiences in adults with relatively mild congenital heart disease. A widespread instrument on subjective health status has previously been applied in studies concerning adults with congenital heart disease (Kamphuis et al., 2002; Lane et al., 2002), though not in a consecutive series with fairly large numbers of adult patients representing specific cardiac diagnostic categories.

Medical predictors

Open heart surgery for congenital heart disease can be considered as a dramatic experience in childhood, both medically as psychologically. Previously, medical variables associated with cardiac surgery, such as circulatory arrest (Bellinger et al., 1997; Utens et al., 1998b) and number of heart operations (Utens et al., 1998b), were identified as predictors for long-term behavioural and emotional problems in children and adolescents with congenital heart disease. To identify patients at risk, it is important to know to what extent factors associated with the medical course in childhood are predictive for long-term behavioural and emotional problems in adulthood. In addition, the effect of the more recent medical course on the patient's psychosocial functioning is to be investigated. Aspects in the treatment and support of adults with congenital heart disease which need special attention can then be specified.

The present study: a 10 year longitudinal cohort follow-up

The study described in this thesis concerns the second follow-up of a longitudinal cohort study. During the first follow-up, conducted in 1989 through 1991, the patients' ages varied from 10 to 35 years. Problems in psychosocial functioning were reported for children and adolescents with congenital heart disease, but not so much for young adult patients (Utens et al., 1998a; Utens et al., 1996; Utens et al., 1994; Utens et al., 1993). At the time of the second follow-up, conducted in 2000 through 2001, all patients of the initial sample had reached adult age, ranging from 20 to 46 years. This offered the opportunity to study the psychosocial functioning of adults with congenital heart disease in a larger sample of only adults, with a wider age range in adulthood. Five large cardiac diagnostic groups were assessed with standardised instruments. During the second follow-up, new topics which are relevant to congenital heart disease in adulthood, such as coping styles, social support and subjective health experiences, were added to the psychological assessment procedure. Moreover, the longitudinal measurements offered an opportunity to describe the long-term development of psychosocial functioning from childhood, adolescence and young adulthood into (later) adulthood. Since the full cohort of patients received their first open heart surgery in the institution where this study was conducted, and extensive medical examination was performed during the first as well as the second follow-up, there was access to a wide range of medical variables. This enabled us to investigate the value of medical variables, derived from the complete medical course from birth until now, to predict emotional and behavioural problems in these adult patients.

Aims

In the present study, various indicators of the psychosocial functioning of adults with congenital heart disease were determined, including biographical characteristics, social functioning, emotional functioning, psychological well-being, psychopathology, coping styles, social support and subjective health experiences. Psychosocial functioning indicators of the patient sample were compared with those in the normal population. Also, the developmental course of some aspects of psychosocial functioning was determined. The role of age, gender and cardiac diagnoses is systematically examined. Furthermore, the predictive value of a wide range of medical variables on emotional and behavioural problems in adults with congenital heart disease is examined.

The aims of the present study were:

1. To compare the present psychosocial functioning of adults with congenital heart disease with that of normative samples.
2. To determine the longitudinal course of psychosocial functioning of adults with congenital heart disease.
3. To determine the role of age, gender and cardiac diagnostic category on psychosocial functioning of adults with congenital heart disease.
4. To determine the predictive value of a wide range of medical variables on emotional and behavioural problems of adults with congenital heart disease.

Methods

Inclusion criteria

During the first follow-up, which took place in 1989-1991, all consecutive patients who underwent their first open heart surgery for congenital heart disease between 1968 and 1980 in the Erasmus MC, and who were younger than 15 years at the time of surgery, were eligible. The total sample of 498 patients then comprised 210 children and adolescents (10-17 years) and 288 adults (18-35 years).

The target population of the present follow-up (2000-2001) consisted of the 498 patients of the first follow-up. From the 498 patients we excluded 61 patients with a variety of congenital heart defects (aortic stenosis, discrete subaortic stenosis, atrioventricular septal defect, pulmonary atresia, tricuspid artresia, total abnormal pulmonary venous drainage, truncus arteriosus, and further miscellaneous diagnoses). Further, we excluded 11 deceased patients, 26 untraceable patients and 1 patient who had undergone a heart transplantation.

Patient sample

Of the remaining 399 patients, 37 refused to participate. The present patient sample consisted of the remaining 362 adult patients (mean age: 30.2 years, age range 20-46 years) of which 194 were males and 168 were females. The overall response rate was 90.7%. For some questionnaires, the response rate was somewhat lower because some were not usable or applicable due to incomplete information, mental retardation or language problems. The numbers are indicated in the corresponding chapters.

The numbers of patients belonging to different diagnostic groups were: atrial septal defect (ASD, N=93), ventricular septal defect (VSD, N=97), tetralogy of Fallot (ToF, N=77), transposition of the great arteries (TGA, N=55) and pulmonary stenosis (PS, N=40). The mean ages in these diagnostic groups were 32.8, 29.5, 30.2, 26.0, and 31.3 years, respectively.

Instruments

During the psychological examination, a semi-structured interview was used to assess psychosocial aspects, such as biographical variables: living conditions, offspring, education, marital and occupational status (Rotterdams Kwaliteit van Leven – Interview; Utens et al., 2000b). Social functioning was assessed with standardised items derived from the Netherlands Central Bureau of Statistics (CBS; Netherlands Central Bureau of Statistics, 1986, 2000). The Dutch Personality Questionnaire (DPQ; Luteijn et al., 1985) was used to assess emotional functioning. Psychological well-being was assessed with the Heart Patients Psychological Questionnaire (HPPQ; Erdman, 1982). Psychopathology was assessed with the Young Adult Self-Report (YASR; Achenbach, 1997) and the Young Adult Behavior Checklist (YABCL; Achenbach, 1997). The Utrecht Coping List (UCL; Schreurs et al., 1993) was used to assess coping styles. Social support was assessed by the Social Support List (SSL; van Sonderen, 1993). Subjective Health status was assessed by the SF-36 (Ware & Sherbourne, 1992). A detailed description of these instruments can be found in the concerning articles.

Assessment procedure

All patients were traced, approached uniformly and signed an informed consent before participating. During their visit to the Erasmus Medical Centre Rotterdam, all patients were interviewed and tested by the same psychologist (EvR) and medically examined by a cardiologist. Some patients (N=28), who could not visit the hospital for practical or emotional reasons, completed questionnaires at home and returned them by mail. The interview then was carried out by telephone. One questionnaire, the Young Adult Behaviour Checklist, was completed by someone familiar to the patient (such as a spouse or parent).

The structure of this thesis

In chapter 2, information is provided on different aspects of psychosocial functioning in adults with congenital heart disease. A wide range of biographical characteristics, concerning living conditions, offspring, education, marital and occupational status is investigated.

Emotional functioning is evaluated in terms of personality traits. Social functioning is described by the patients' participation in leisure-time activities. In chapter 3, the present status of psychopathology as well as the longitudinal development over a 10 year period is investigated. Differences in sex and age between different groups who show different patterns as to the development of psychopathology are investigated. In chapter 4, coping styles in adults with congenital heart disease and the social support they receive are examined. The association between coping styles and social support in the patient sample is investigated. In chapter 5, the present subjective health status is assessed by an internationally widespread instrument. Also, the longitudinal course of psychological well-being from follow-up 1 to follow-up 2 is examined with an instrument that was especially designed for heart patients. In chapter 6, medical predictors, covering the medial course from birth up till the present adult life, for long-term behavioural and emotional problems in adulthood, are identified. Finally, in chapter 7, the main findings and conclusions of this thesis are discussed. Implications and recommendations for medical practise are given.

2 | Psychosocial functioning of the adult with congenital heart disease: a 20-33 years follow-up

European Heart Journal

2003, 24: 673-683

E.H.M. van Rijen

E.M.W.J. Utens

J.W. Roos-Hesselink

F.J. Meijboom

R.T. van Domburg

J.R.T.C. Roelandt

A.J.J.C. Bogers

F.C. Verhulst

Chapter 2

Psychosocial functioning of the adult with congenital heart disease: a 20-33 years follow-up

Abstract

Aims: *Since knowledge about the psychosocial function of adult patients with congenital heart disease is limited, we compared biographical characteristics, and emotional and social functioning of these patients with that of the reference groups.* ***Methods and results:*** *Patients with congenital heart disease (N=362, aged 20-46 years), belonging to five diagnostic groups, were subjected to extensive medical and psychological examination, 20-33 years after their first open heart surgery. All the patients were seen by the same psychologist, who examined their psychosocial functioning using a structured interview and questionnaires. The majority (78%) was living independently and showed favourable outcome regarding the marital status. Among married/ cohabitant patients, 25-39-year-olds showed normal offspring rates. None of the 20-24-year-old patients had any children. The offspring rate dropped after the age of 40. The proportion of adult patients with a history of special education was high (27%). Accordingly, patients showed lower educational and occupational levels compared to reference groups. As regard to the emotional and social functioning (leisure-time activities), the sample showed favourable results.* ***Conclusions:*** *Overall, this sample of patients with congenital heart disease seemed capable of leading normal lives and seemed motivated to make good use of their abilities.*

Introduction

With the improvement of surgical techniques over the last decades, increasing survival rates of patients with congenital heart disease are resulting in a new and growing patient population: adults with operated congenital heart disease. Medical professionals not only encounter specific medical needs of these patients, but also deal with unique psychosocial characteristics of adults with congenital heart disease. So far, however, little is known about the specific psychosocial problems of these patients when they enter adulthood, or the extent to which they succeed in having a normal life. This might impede medical professionals in tailoring treatment to the specific needs of adult patients with congenital heart disease, since these patients differ in many aspects from the well-known adult population with acquired cardiac diseases. The purpose of this study was to provide a clear and solid view on the biographical characteristics and current emotional and social functioning of the adults with congenital heart disease.

Both positive and negative outcomes have been documented regarding the educational and occupational functioning (Brandhagen et al., 1991; Gersony et al., 1993; Kokkonen & Paavilainen, 1992; Shampaine et al., 1990; Ternestedt et al., 2001; Utens et al., 1994), living conditions, marital status and offspring (Gersony et al., 1993; Kokkonen & Paavilainen, 1992; Ternestedt et al., 2001; Utens et al., 1994) and emotional and social functioning (Brandhagen et al., 1991; Shampaine et al., 1990; Ternestedt et al., 2001; Utens et al., 1994). Deriving a complete picture of the adult patient with congenital heart disease from the existing studies is hampered by several factors, including small sample size, heterogeneous sample composition with regard to the type of congenital heart disease, different age ranges, low-response rate, non-standardised assessment procedures and variation in methods across various studies.

This study is part of a multidisciplinary cohort study of patients with congenital heart disease, 20-33 years after their first heart surgery. This work concerns the second follow-up of the cohort and offers the unique possibility to study its current psychosocial functioning with a larger sample size and age range in adulthood compared to previous works. It has been suggested that patients rate the negative impact of the congenital heart disease on their functioning as more serious as they grow older (Ternestedt et al., 2001). However, very little information is available about how these patients function later in adulthood.

Therefore, the aims of the present study were (1) to compare the biographical characteristics and emotional and social functioning of adults with operated congenital heart disease with that of reference groups; (2) to determine the relationship between cardiac diagnosis and the biographical characteristics and emotional and social functioning of the patients; and (3) to determine the effects of sex and age on biographical characteristics and on emotional and social functioning.

Methods

Inclusion Criteria

During the first follow-up of this study, which took place between 1989 and 1991, all consecutive patients who underwent their first open heart surgery for congenital heart disease between 1968 and 1980 in the Erasmus MC, and who were younger than 15 years at the time of surgery, were eligible. This population is described in detail elsewhere (Utens et al., 1994).

The target population of the second follow-up (2000-2001) consisted of the 498 patients of the first follow-up. From the 498 patients we excluded: 11 deceased patients (one with atrial septal defect, four with ventricular septal defect, two with tetralogy of Fallot, two with transposition of the great arteries and two with pulmonary stenosis), 26 untraceable patients and one patient who had undergone heart transplantation; 61 patients were also excluded because they belonged to a miscellaneous diagnostic group, consisting of a small number of patients with a variety of congenital heart defects.

Patient sample

Of the remaining 399 eligible patients, 37 refused to participate. The present patient sample consisted of the remaining 362 adult patients (mean age: 30.2 years, age range 20-46 years), of which 194 were males and 168 were females. For biographical characteristics, data were available for all the patients, but not on all topics. For emotional functioning, data were missing for 11 patients (eight were mentally retarded, one had a linguistic problem, two did not complete the questionnaire). For social functioning, data were missing for two persons (one was mentally retarded, one did not complete the questionnaire). The total number of patients answering each question(naire) are indicated in Tables 2.1-2.5. The overall response rate, corrected for deceased patients and persons lost to follow-up, was 90.7%.

The total number of patients belonging to different diagnostic groups was: atrial septal defect (ASD, N=93), ventricular septal defect (VSD, N=97), tetralogy of Fallot (ToF, N=77), transposition of the great arteries (TGA, N=55) and pulmonary stenosis (PS, N=40). The mean ages in these diagnostic groups were 32.8, 29.5, 30.2, 26.0 and 31.3 years, respectively. Medical history and physical condition of these patients at the time of the first follow-up are described elsewhere (Meijboom et al., 1993; Meijboom et al., 1995; Meijboom et al., 1996; Meijboom et al., 1994).

For the biographical characteristics and social functioning, recent normative data were derived from the Netherlands Central Bureau of Statistics and, wherever available, were specified by age and sex (Netherlands Central Bureau of Statistics, 2002). These recent reference data were derived from a variety of normative samples. Since these concerned very large samples, the representativeness for the average Dutch situation was warranted. Sample sizes of multiple and very large reference groups were not indicated in the Tables 2.1-2.5 to prevent confusion. For emotional functioning, the reference group consisted of 5686 adults (mean age, 34.6 years, age range 15-65 years; 2730 males and 2956 females), derived from an extensive Dutch population study (Luteijn et al., 1985).

Instruments

Biographical characteristics. A structured interview was designed to assess biographical variables, such as living conditions, offspring, education and marital and occupational status. Special education included schools for learning-disabled, mentally handicapped and chronically ill children.

Emotional functioning. From the Dutch Personality Questionnaire (DPQ; Luteijn et al., 1985), derived from the California Psychological Inventory (Gough, 1964), three scales were used, namely Hostility (19 items), Self-esteem (19 items) and Neuroticism (21 items). Hostility measures the extent of criticism, distrust and intolerance towards other people. Self-esteem measures a positive attitude towards work, flexibility and being energetic and self-controlled. Neuroticism measures feelings of stress, depression, unstableness and insecurity. The response possibilities of the Dutch Personality Questionnaire are: yes = 2, do not know = 1, and no = 0. A high score of self-esteem indicates a high self-esteem. On the other scales, the higher the scores, the poorer the social adjustment. The internal consistency, stability and validity of the DPQ can be considered favourable (Luteijn et al., 1975; Luteijn et al., 1985). The psychometric qualities of the DPQ are described elsewhere in more detail (Utens et al., 1994).

Social functioning. Leisure-time activities were assessed with standardised items derived from the Netherlands Central Bureau of Statistics (Netherlands Central Bureau of Statistics, 1986, 2000).

Assessment procedure

During their visit to the Erasmus Medical Centre, patients were interviewed and tested by a psychologist (EvR) and medically examined by a cardiologist. For patients with low intellectual functioning, the question(naire)s were verbally administered, if possible. Of the 362 patients, 28 patients preferred to complete the questionnaires at home. In these cases, the structured interview was done by telephone, except for five patients who refused such an interview.

Statistical analyses

For biographical characteristics, proportions of patients are presented in percentages and actual numbers. Levels of significance were calculated only when the patient sample was comparable to reference groups with regard to age and gender. In order to correct for multiple comparisons, only the differences in biographical characteristics with a level of significance lower than 0.01 were considered significant. As to the emotional and social functioning, for both the patient sample and for the reference groups, 95% confidence intervals (CI) were calculated, respectively, around the group means of the DPQ scales and the proportions on leisure-time activities. If the 95% CI of the patient sample did not overlap with those of the reference group, the differences between the group means or the participating proportions were considered significant. Comparisons between diagnostic categories were also performed based on 95% CI. Owing to small cell sizes, statistical differences between diagnostic categories could not always be proven. For the DPQ, Cohen's Ds (Cohen, 1988) were computed to assess the magnitude of differences in mean scores between the patient sample and the reference groups. According to Cohen's (1988) criteria, a standardised difference of 0.20 can be considered as small, 0.50 as medium, and 0.90 as high.

Results

Biographical characteristics

The main biographical characteristics are outlined in Table 2.1. The majority of the patient sample (78%) was living on their own (defined as living independently), while a total of 18% was not living independently, but with parents or another care-taker. When specified for age and sex categories, the living conditions of the patient sample showed high similarities with those of the reference group. The 25-29-year-old females from the patient sample were living on their own, significantly more often compared to the reference group ($p<0.01$). Patients appeared to be living in (or under supervision of) institutions for mentally handicapped significantly more often than the reference group (4% vs. 1%; $p<0.01$), with no significant differences between diagnostic categories.

With respect to the marital status of the patient sample, at least 72% was in some sort of a relationship at the time of the interview. For persons living independently (N=279), no significant differences in marital status were found between the patient sample and the reference group.

Regarding the offspring, a total of 34% of the patient sample had one or more child(ren). Among married persons or cohabitants (N=217), none of the 20-24-year-old patients had any children, in contrast to 14% in the reference group. Within the age of 25-39 years, no significant differences in offspring were found between the patient sample and the reference group. The proportion of patients with congenital heart disease with offspring drops after the age of 40. Based on median split age groups, no significant differences between the diagnostic categories were found.

Table 2.1 Biographical characteristics of the patient sample

	ASD (%)	VSD (%)	ToF (%)	TGA (%)	PS (%)	Total patient sample (%)
	(N=93)	(N=97)	(N=77)	(N=55)	(N=40)	(N=362)
Living conditions (N=357)						
With parents	6	18	20	39	10	17 (62)
On one's one	92	77	75	57	82	78 (279)
In institution for mentally handicapped	2	5	5	4	5	4 (15)
Other	0	0	0	0	3	< 1 (1)
Marital status (N=357)						
No relationship	21	26	25	38	22	26 (93)
Stable relationship	6	16	12	17	2	11 (39)
Cohabitant	16	19	24	17	28	20 (71)
Married	53	35	37	26	48	40 (143)
Divorced	3	3	1	2	0	2 (8)
Cohabitant or married after divorce	1	1	1	0	0	1 (3)
Offspring (N=357)						
0 children	54	72	63	85	55	66 (234)
1 child	12	12	13	9	18	12 (44)
2 children	24	12	16	4	20	16 (56)
≥ 3 children	10	4	8	2	7	6 (23)
Former course of education (N=358)						
Special education	13	28	33	40	23	27 (95)
Regular education	87	72	67	60	77	73 (263)
Educational attainment (N=334)						
Lower	47	43	52	48	43	47 (156)
Average	31	36	32	31	43	34 (114)
Higher	22	21	16	21	14	19 (64)
Daily activities (N=362)						
Attending school	2	5	7	9	0	5 (18)
Job	83	82	73	71	77	78 (282)
Unemployed[a]	0	0	3	7	0	2 (6)
Disablement pension[a]	1	0	3	4	0	1 (5)
Volunteer, unpaid work[a]	2	3	1	2	0	2 (7)
Long-term sick leave	2	1	0	2	2	1 (5)
Household	6	1	3	0	10	3 (12)
Labour institution for mentally handicapped	4	6	5	5	8	6 (20)
Activity centre for mentally handicapped[a]	0	2	5	0	3	2 (7)

Note. Abbreviations: ASD, atrial septal defect; VSD, ventricular septal defect; ToF, tetralogy of Fallot; TGA, transposition of the great arteries; PS, pulmonary stenosis. Between parentheses, the actual numbers of patients for which data were available are indicated.

[a] Considered to receive social security benefits.

Table 2.1 shows that 27% of the patient sample had followed some sort of special education in the past. Of these patients, 85% attended schools for learning-disabled or mentally handicapped children and 15% for chronically ill children. The proportions of patients with a history of special education were significantly higher in the diagnostic categories tetralogy of Fallot (33% (22-44)) and transposition of the great arteries (40% (27-53)) than in the atrial septal defect group (13% (6-20)).

The educational attainments of the patient sample were evaluated excluding those living in institutions for mentally handicapped. For the patient sample, the highest educational level completed was significantly more often a lower level in comparison to the reference group (47% vs. 39%, $p<0.01$). No significant differences in educational attainments were found between the diagnostic categories.

Regarding the daily activities of the patient sample, at least 85% had a paid job (persons on long-term sick leave and working in labour institutions for mentally handicapped included), 7% received some sort of social security benefit and 8% were either attending school or doing full-time household. Normative data on daily activities were not available. Although no significant differences between diagnostic categories were found, social security benefits were received most often in the diagnostic categories tetralogy of Fallot and transposition of the great arteries (12% and 13% respectively).

The occupational status of the patient sample is further outlined in Table 2.2. The patient sample appeared to be significantly overrepresented in the lower level occupations, and underrepresented in the scientific occupations, compared to the reference group. No significant differences in occupational status were found between the diagnostic categories. As to the duration of employment, the patient sample did not differ from the reference group, nor were there any significant differences between diagnostic categories. In the decision to work part-time, the congenital heart disease played a role in 21% of the cases. Although no statistical differences between diagnostic groups were revealed, the congenital heart disease was mentioned as a reason to work part-time most often in the diagnostic categories, transposition of the great arteries, pulmonary stenosis, and tetralogy of Fallot (40%, 36% and 29% respectively). Besides specific cardiac problems, weariness was often mentioned as another reason. The estimation of the gross income in the patient sample exceeded that of the reference group to some extent. The congenital heart disease sick-leave percentage was higher than that of the reference group. This was not in accordance with their own perception, since

Table 2.2 Occupational status of persons with paid work in the patient sample and reference group (persons in labour institutions included)

	Patient sample (N=302)			Reference group		P-value
	% (N)	M	SD	%	M	
Occupational level (SBC 1992)[a] (N=236)[b]						
Elementary	5 (11)			6		ns
Lower	34 (79)			22		< 0.001
Average	39 (93)			41		ns
Higher	18 (43)			22		ns
Scientific	4 (10)			9		< 0.001
Duration of employment (N=295)						
Full-time (≥ 36 hours a week)						
Total	69 (205)			70		ns
Male	91 (152)			89		ns
Female	41 (53)			40		ns
Part-time						
Total	31 (90)			30		ns
Male	9 (15)			11		ns
Female	59 (75)			60		ns
Reason part-time employment						
Unrelated to heart disease	79 (71)					
Heart disease one of several reasons	11 (10)					
Heart disease the only reason	10 (9)					
Income (gross salary divided by 1000 euros) (N=220)						
Total		25.9	18.7		21.0	
Sick leave (from previous year) (N=297)[c]						
Mean sick-leave percentage[d]		7.9	31.3		5.5	
Sick leave according to patients						
Less than colleagues	57 (162)					
Equal to colleagues	30 (86)					
More than colleagues	13 (36)					
Reason sick leave						
Unrelated to heart disease	92 (169)					
Heart disease one of several reasons	4 (7)					
Heart disease the only reason	4 (7)					
Perception of career possibilities (N=285)						
Career possibilities according to patients						
Equal to colleagues	92 (262)					
Less than colleagues	8 (23)					

Note. Between parentheses, the actual numbers of patients for which data were available are indicated.

[a] Standaard Beroepen Codering 1992 (Standard Occupation Classification), Netherlands Central Bureau of Statistics.

[b] Only persons ≥ 25 years are included, persons living in institutions for mentally handicapped are excluded.

[c] Persons on long-term sick leave are also included in this section (total N=307).

[d] Sick-leave percentage was used in stead of number of days on sick leave, since the number of hours a person works a week (full-time/ part-time) might vary considerably.

the majority of the patient sample reported to have a lower sick leave than colleagues. Although no statistical differences were revealed, the sick-leave percentage was highest in the diagnostic categories, tetralogy of Fallot and transposition of the great arteries (10.7% and 10.1% respectively). The congenital heart disease was mentioned as a reason for sick leave in 8% of all cases. Among the diagnostic categories, this was the case most often within the category, transposition of the great arteries (18%). The patients' estimation of career possibilities seemed favourable and equal for all the diagnostic categories. Table 2.3 provides an overview of the main, more objective, occupational variables for the five different diagnostic categories.

Table 2.3 Summary of main variables concerning occupational status for different cardiac diagnostic groups

	ASD		VSD		ToF		TGA		PS	
	%	M	%	M	%	M	%	M	%	M
Occupational level (SBC 1992)[a] (N=236)[b]										
Elementary	5		6		2		0		7	
Lower	31		35		47		28		19	
Average	42		30		34		48		55	
Higher	18		22		13		19		19	
Scientific	4		7		4		5		0	
Duration of employment (N=295)										
Full-time (≥ 36 hours a week)	68		68		71		75		68	
Part-time	32		32		29		25		32	
Income (gross salary divided by 1000 euros) (N = 220)		27.5		25.0		24.4		21.5		32.9
Sick leave (from previous year) (N=297)[c]										
Mean sick-leave percentage[d]		6.3		8.5		10.7		10.1		2.6

Note. Abbreviations: ASD, atrial septal defect; VSD, ventricular septal defect; ToF, tetralogy of Fallot; TGA, transposition of the great arteries; PS, pulmonary stenosis.

[a] Standaard Beroepen Codering 1992 (Standard Occupation Classification), Netherlands Central Bureau of Statistics.

[b] Only persons ≥ 25 years are included, persons living in institutions for mentally handicapped are excluded.

[c] Persons on long-term sick leave are also included in this section (total N=307).

[d] Sick-leave percentage was used instead of number of days on sick leave, since the number of hours a person works a week (full-time/ part-time) might vary considerably.

Emotional functioning

On the DPQ, the patient sample obtained significantly more favourable results on hostility, self-esteem and neuroticism than the reference group (Table 2.4). The differences in mean scores could all be considered as medium, according to Cohen's (1988) criteria, except for the effect, for females, on neuroticism, which could be considered as small.

Of the 351 patients who completed the DPQ, 18 patients fulfilled their daily activities in either a labour institution or an activity centre for mentally handicapped and 25 patients completed it at home (one patient belonged to both of these groups). It was assumed that the DPQ reference group did not contain any mentally handicapped. Furthermore, possible bias from home-completed questionnaires should be excluded. Therefore, DPQ scores for the patient sample, after excluding those visiting labour institutions or activity centres for mentally handicapped and patients who completed the DPQ at home (DPQ-Excluded) were calculated. The results of the DPQ-Excluded group were only slightly different from those of the original 351 patient-respondents (Table 2.4). The DPQ-Excluded group also obtained significantly more favourable results on hostility, self-esteem and neuroticism than the reference group (Cohen, 1988).

Table 2.4 Mean scores, 95% CI, standard deviations and Cohen's D on the DPQ[a] for patient sample and reference group and DPQ-EX.[b] group

DPQ[a]	Patient sample (N=351)		Reference (N=5686)		Cohen's D
	M (CI)	SD	M (CI)	SD	
Hostility	14.9 (14.1-15.6)	7.2	18.2 (18.0-18.4)	6.7	0.5
Self-esteem	31.0 (30.5-31.6)	5.3	28.0 (27.9-28.1)	5.6	0.5
Neuroticism					
Male[c]	6.5 (5.6- 7.4)	6.3	10.1 (10.0-10.2)	7.5	0.5
Female[c]	10.3 (9.2-11.5)	7.6	13.9 (13.6-14.2)	8.3	0.4
DPQ-EX. (N=307)					
Hostility	15.0 (14.2-15.8)	7.2			0.5
Self-esteem	31.2 (30.5-31.8)	5.4			0.6
Neuroticism					
Male[d]	6.5 (5.5- 7.4)	6.4			0.5
Female[d]	10.2 (8.9-11.4)	7.5			0.4

[a] Dutch Personality Questionnaire (Luteijn et al., 1985).
[b] DPQ-EX.: Dutch Personality Questionnaire-results of patients after exclusion of mailed questionnaire data and patients attending either labour institutions or activity centres for mentally handicapped.
[c] Male: patients N=190, reference N=2730; female: patients N=161, reference N=2956.
[d] Male: patients N=167; female: patients N=140.

Effects of diagnostic category, sex and age on emotional functioning of the patients were computed on the DPQ scales. No significant differences were found between the diagnostic categories. As for sex effects, females scored significantly less favourable on self-esteem compared to males (females: mean = 29.9 (29.0-30.8); males: mean = 32.0 (31.3-

32.7)). Furthermore, in accordance with the reference group, females reported significantly more complaints on neuroticism than males (females: mean = 10.3 (9.2-11.5); males: mean = 6.5 (5.6-7.4)). To assess age effects, two categories (20-29 years and 30-46 years), based on the median split of the patient sample, were formed. No significant age effect was found.

Since the numbers of male vs. female and younger vs. older patients differed across the diagnostic categories on all DPQ scales, sex and age effects were next tested when adjusted to diagnostic category. For each diagnostic category, 95% CI around the mean scores on the DPQ scales of male vs. female, and younger vs. older patients were compared. No significant sex or age effects were found.

Social functioning

Table 2.5 presents significant differences in participation in leisure-time activities between the patient sample and reference group, for different age- and sex-based groups. For all of these differences, participation was higher in the patient sample than in the reference group. Higher participation in leisure-time activities was reported for outgoing activities like going to the movies, visiting clubs and discos and shopping, as well as for domestic activities like playing games and watching TV.

When total sport participation ($\geq$ 1 hour a week) between the diagnostic categories was compared, no significant differences were found. More intensive sport participation ($\geq$ 5 hours a week), however, was found significantly more often in the diagnostic category, ventricular septal defect than in the transposition of the great arteries group (19% (11-27) and 4% (0-9), respectively). Apart for brain-teasers, which were performed significantly more often by patients with ventricular septal defect and transposition of the great arteries than by patients with tetralogy of Fallot and pulmonary stenosis, no further significant differences in participation in leisure-time activities between diagnostic categories were found.

Table 2.5 Significant differences in leisure-time activities (proportions of participation and 95% CI) between patient sample and reference group

Item	18-24 years				25-34 years				35-44 years			
	Males		Females		Males		Females		Males		Females	
	Patient sample (N=50)	Refe-rence group	Patient sample (N=27)	Refe-rence group	Patient sample (N=100)	Refe-rence group	Patient sample (N=98)	Refe-rence group	Patient sample (N=43)	Refe-rence group	Patient sample (N=40)	Refe-rence group
	% (CI)	% (CI)	% (CI)	% (CI)	% (CI)	% (CI)	% (CI)	% (CI)	% (CI)	% (CI)	% (CI)	% (CI)
Movies							69 (60–79)	53 (48–58)	60 (45–76)	36 (31–41)		
Playing games									88 (78–98)	66 (61–71)	93 (84-100)	73 (68–78)
Walking/ cycling			100	95 (93–97)								
Visiting clubs					49 (39–59)	32 (28–36)			53 (38–69)	24 (19–29)		
Visiting discos							43 (33–53)	25 (21–29)				
Watching TV											100	95 (93–97)
Shopping					79 (71–87)	54 (49–59)	96 (92-100)	83 (79–87)	77 (64–90)	49 (44–54)	95 (88–100)	77 (73–81)

Note. Only significant differences were reported in the table. No significant differences were shown on the following items not reported in the table: watching sports, brain-teasers, making music/acting, handicrafts, odd jobs, listening to radio, doing sports, visiting bars.

Discussion

Biographical, emotional and social features of the patient sample

Overall, living conditions, marital status and offspring of the patient sample in the present study showed high similarity with the reference groups. Regarding living conditions, Ternestedt et al. (2001) also found similarities between patients with congenital heart disease and a normative sample. Kokkonen & Paavilainen (1992) and Utens et al. (1994), however, did find differences: especially the young patients with congenital heart disease were living with their parents more often than reference groups. The higher mean age and larger age range in adulthood in the present patient sample, compared to the relative young samples of the previous studies (Kokkonen & Paavilainen, 1992; Utens et al., 1994) provide a possible explanation for these diverging findings. The previously found arrears in living conditions might be equalised later on in adulthood. As to marital status, the results of this present study confirm to those of Ternestedt et al. (2001), but are in contrast with the results of Kokkonen & Paavilainen (1992) and Gersony et al. (1993). The latter found patients with congenital heart disease to be married less often, or get married at a later age compared to the reference groups. It should, however, be mentioned that in the present study, marital status was scrutinised for a selection of the patient sample, namely those already living independently. Offspring was examined only for married persons or cohabitants. Furthermore, in this study, both sexes were included, instead of only females, which might explain the favourable outcome in offspring in contrast to Gersony et al. (1993), who found elevated childlessness among congenital heart disease women. It should be noted that decreased rates of offspring for the youngest (20-24 years) and oldest (above 40 years) patients of the sample could not be proven significantly different because of empty and small cells.

In the present study, the proportion of patients with a history of special education can be considered as high. For 15% of those the impact of the congenital heart disease on their school career (hospitalisation, restrictions) seemed to be the main reason for attendance of special education, since they had been in schools for chronically ill children. In the remaining 85%, some sort of learning-disability or mental retardation seemed apparent, considering the type of special education they had attended. Additionally, within the patient sample, the highest educational level completed was more often a lower level compared to the reference group. Although it is well known that congenital heart disease patients encounter different barriers in their educational course during childhood, such as absence from school due to illness, treatment or recovery (Casey et al., 1994; Wray & Sensky, 2001) and learning-disabilities (Bellinger et al., 1999; Ellerbeck et al., 1998; Wray & Sensky, 2001), the high number of adult patients with congenital heart disease with a history of special education never appeared so clearly as in this study. This high attendance of special education in the past might have resulted in lower educational attainments in adulthood: a finding, which is highly in contrast with the findings of Brandhagen et al. (1991) and Ternestedt et al. (2001) who found an even higher educational level in the adults with congenital heart disease compared to the normative groups.

In accordance with the educational attainments, this patient sample had lower level of occupations significantly more often than the reference group. The duration of employment of the patient sample, however, did not differ from the reference group. The gross income of the patient sample seemed somewhat higher than the reference group. However, it should be mentioned that most patients could only give a rough indication of their income and, owing to social desirability, they might have exaggerated their estimations. The sick-leave percentage of the patient sample was higher than that of the reference group, although most patients estimated that their sick leave was less than colleagues. This might indicate possible denial. Accordingly, patients with congenital heart disease seemed to have a positive perception of their career possibilities. Overall, it can be stated that, despite a somewhat lower occupational level and higher sick leave, the present patient sample showed favourable results regarding the duration of employment, income and perception of career possibilities. Broadly speaking, this confirms the positive findings of previous studies (Gersony et al., 1993; Kokkonen & Paavilainen, 1992; Shampaine et al., 1990) with respect to employment.

Regarding emotional functioning, Brandhagen et al. (1991) found an increased level of psychological stress in adults with congenital heart disease. Shampaine et al. (1990) found both positive and negative emotional outcomes. Ternestedt et al. (2001) found more favourable emotional outcomes in patients with tetralogy of Fallot compared to patients with atrial septal defect. The present patient sample showed more favourable scores than the reference group on hostility, self-esteem and neuroticism. These findings are similar to those of Utens et al. (1994), who studied the same cohort with the same instrument regarding emotional functioning 10 years earlier. In this study (Utens et al., 1994), it was suggested that denial mechanisms and high achievement motivation possibly lead congenital heart disease patients to overrate their emotional states. Recently, perseverance has been put forward as a coping mechanism in congenital heart disease patients (Ternestedt et al., 2001).

Social functioning of the patient sample can be considered favourable. If significant differences were found in participation in leisure-time activities, the congenital heart disease patients obtained more positive scores than the reference group. These overall favourable results on social functioning confirm earlier favourable findings (Gersony et al., 1993; Utens et al., 1994).

Effect of cardiac diagnosis

Regarding biographical characteristics, no significant differences between the diagnostic categories were found for living in institutions, having offspring, educational attainments, receiving social security benefits, occupational level and duration of employment. Regarding social functioning, participation in leisure-time activities was highly similar among the different diagnostic categories, except for intensive sport participation.

Overall, it can be stated that individuals within the diagnostic categories transposition of the great arteries and tetralogy of Fallot showed some residual effects from the congenital heart defect as to participation in education, occupation and intensive sport, but further were able to live normal lives. This finding is supported by the fact that no significant differences in emotional functioning were found between the diagnostic categories.

Effects of sex and age

As to living conditions, the 25-29-year-old female patients were living on their own significantly more often than the reference group. Regarding the offspring, the patients seemed to raise families somewhat later in their 20s, when compared with the reference group. Remarkably, there was a drop of patients with offspring after the age of 40. This is probably due to a cohort effect since for the eldest of the patient sample, the cardiac surgery was not yet available when they were born. Therefore, patients were more likely to be operated at an older age and treated with less advanced techniques compared to the youngest patients in this sample. Also, they were the first congenital heart disease patients to enter adulthood and experience uncertainties regarding having offspring. The impact of the congenital heart disease on having offspring may, therefore, be greater for the older patients in the sample, compared to the younger ones.

With respect to emotional functioning, female patients scored significantly less favourable on neuroticism (in accordance with the reference group) and self-esteem than male patients. This is consistent with previous findings (Utens et al., 1998a), which showed that females with congenital heart disease report more somatic complaints than males with congenital heart disease.

Limitations

It should be noted that the present patient sample contains a selection of five frequent diagnostic categories and may, therefore, not be completely representative of all the congenital heart disease anomalies. The present patient sample concerns a generation of congenital heart disease patients who were treated with different techniques than the ones used at present. Since major advances took place in surgical techniques and psychosocial needs could have changed, the results of this study might not fully apply to young patients with congenital heart disease operated upon these days. Biographical characteristics and social functioning of the present study's sample might not be directly applicable to other countries and cultures, since life styles or aspects, for example, availability of social security or educational systems, might differ considerably. Therefore, caution should be taken in generalising the psychosocial functioning of patients with congenital heart disease as described in this study.

For biographical variables, normative data were not always available. Some biographical variables were only looked at under selected conditions (marital status of independently living persons and offspring of cohabitants or married persons). This might have attributed to the favourable results on these variables for the patient sample. Furthermore, some diagnostic categories had relatively small sample sizes, which implied larger 95% CI for these groups. This may have hampered finding differences between diagnostic groups. Taking into account the large amount of comparisons that were made in this study, the amount of differences in psychosocial functioning between the patient sample and reference group can be considered limited. This strengthens the overall positive outcome for this sample of adult patients with congenital heart disease on biographical characteristics, emotional and social functioning.

Implications

The present study shows that this sample of adult patients with congenital heart disease has residual problems regarding its educational and occupational status, which seems a reflection of disadvantaged positions in childhood as can be concluded from the high attendance of special education in the past. However, patients seem to make good use of their abilities as can be concluded from their favourable outcomes on duration of employment and income, and their optimistic outlook on career possibilities. The diagnostic category transposition of the great arteries seems to be hampered by the congenital heart disease most often in their employment, since they mentioned it most often as a reason for part-time working or sick leave.

Taking in consideration further results on biographical characteristics, emotional and social functioning, these adult patients seem capable of leading normal lives. However, some precaution must be taken into account. Some of these positive outcomes, for example regarding sick leave, might be influenced by a tendency in these adult patients to give social desirable answers. Whether this should be explained by denial of restrictions or a coping strategy to keep an optimistic view on their possibilities is unclear. Future research should therefore aim at coping strategies in adult congenital heart disease patients, in order to get a better understanding of how they experience their abilities. Further, the longitudinal development of psychosocial functioning of these patients as well as the influence of medical variables (not yet systematically available) should also be the focus of future research.

3 | Development of psychopathology over a 10 year period in a cohort of adults with congenital heart disease

E.H.M. van Rijen

E.M.W.J. Utens

J.W. Roos-Hesselink

F.J. Meijboom

R.T. van Domburg

J.R.T.C. Roelandt

A.J.J.C. Bogers

F.C. Verhulst

Chapter 3

Development of psychopathology over a 10 year period in a cohort of adults with congenital heart disease

Abstract

Background: *Little is known about the development of psychopathology in children with congenital heart disease (ConHD) from childhood into adulthood. In this study, the course of psychopathology in a cohort of ConHD adults, who received their first heart surgery in childhood, was determined longitudinally over a 10-year-period.* ***Methods:*** *At both the first (1989-1991) and second (2000-2001) follow-up, patients (n=251, aged 20-32 years) completed questionnaires during a psychological examination. Parallel instruments were used to measure psychopathology in childhood, adolescence and adulthood.* ***Results:*** *Especially young ConHD females showed higher levels of psychopathology compared to the reference group. Larger differences in psychopathology were found between younger (20-27 years) and older (28-32 years) patients compared to the reference group; the younger patients showed more psychopathology than the older patients. Also, larger differences in psychopathology were found between male and female patients compared to the reference group; female patients showed more psychopathology than male patients. Parents and partners of patients evaluated the patients' behavior as more deviant from the norm than patients themselves. There was a stronger decline of psychopathology in ConHD patients with increasing age compared to reference groups. Female and younger patients showed more variability in trajectories of psychopathology than did male and older patients.* ***Conclusions:*** *Assessment and treatment of psychopathology at young age is highly desirable in ConHD patients. Young adult ConHD females deserve special attention since they might be hampered by disease-specific uncertainties.*

Introduction

Surgical intervention for congenital heart disease (ConHD) is possible since the late 1950s. Today, due to increased survival rates for ConHD patients, health care professionals are dealing with a new and growing group of patients, namely adults with ConHD. In order to meet the needs of this overall first generation of surviving adults with ConHD, more knowledge about the behavioral and emotional problems in this group should be provided.

Children and adolescents with ConHD are reported to be at a higher risk for behavioral and emotional problems in comparison with peers from reference groups. Previous studies reported increased feelings of anxiety and inferiority, elevated impulsiveness (Kramer et al., 1989), elevated levels of emotional and behavioral problems (Bellinger et al., 1997; Utens et al., 1993) and more medical fears (Gupta et al., 1998) in children and adolescents with ConHD compared to normative groups.

Unfortunately, very little is known about how behavioral and emotional problems of children with ConHD develop into adulthood. In a 25-year follow-up Brandhagen et al. (1991) found more psychological distress in a cohort of adults with ConHD than in a reference group. Utens et al. (1998a) found little differences between mean problems scores of a sample of ConHD adults and a normative group. Differences found were limited to only two specific problem areas, namely somatic complaints and strange thoughts and behaviors. In both studies, no relation was found between the severity of the cardiac defect and the level of psychopathology. Cox et al. (2002) found a low prevalence of psychopathology in ConHD adults, using orthopedic patients as a reference group. Differences in sample size, sample composition with regard to the type of ConHD, response rate and assessment procedures might explain these diverging findings. Although some studies identified predictive factors that determine long-term psychosocial wellbeing in ConHD patients, such as maternal anxiety and pampering (Linde et al., 1966), age at time of operation (Baer et al., 1984), number of heart operations and deep hypothermic circulatory arrest (Utens et al., 1998a), no earlier follow-up study has been performed using repeated measurements from childhood- into adulthood to examine the individual course of psychopathology in ConHD adults.

This study is part of a multidisciplinary study which provides longitudinal psychological and medical data, of patients with ConHD, 20 to 32 years after their first heart operation. At the first follow up, which took place in 1989-1991, children and adolescents with ConHD showed significantly more behavioral and emotional problems than peers from reference groups, at least nine years after cardiac surgery (Utens et al., 1993). For the present follow-up study, 20- to 32-year-olds of the five largest diagnostic groups of the same cohort of (meanwhile all adult) patients with ConHD were examined concerning emotional and behavioral problems, using instruments parallel to those of the first follow-up. The present status, as well as the continuity and change of psychopathology, as reported by patients themselves and by a significant other, will be described.

The aims of this study are:

1. To compare the level of emotional and behavioral problems of adult (20 to 32 years) patients operated for ConHD with that of a normative group of similar age.
2. To determine the development of psychopathology in patients operated for ConHD from childhood and adolescence into adulthood and from early adulthood into later adulthood.
3. To identify the role of cardiac diagnosis, sex, age and different informants in this process.

Methods

Description of the samples

Inclusion criteria. During the first follow-up (Time 1) of this study, which took place in 1989-1991, all consecutive patients who underwent their first open heart surgery for congenital heart disease between 1968 and 1980 in the Erasmus Medical Center Rotterdam, and who were younger than 15 years at the time of surgery, were eligible. This population is described in detail elsewhere (Utens et al., 1993).

The present follow-up (Time 2) took place in 2000-2001. The target population consisted of the 498 patients of the first follow-up. From the 498 patients, we excluded 61 patients who belonged to a miscellaneous diagnostic group, consisting of small numbers of patients with a variety of congenital heart defects. Further, we excluded 11 deceased patients, 26 untraceable patients and 1 patient who had undergone a heart transplantation.

Patient sample. Since normative data were missing for patients above 32 years, only 275 eligible patients aged 20-32 years were included, of which 24 (8.7%) refused to participate. The present study focuses on the remaining 251 ConHD patients (mean age: 27 years).

Of the 251 participating ConHD patients, 8 mentally retarded patients were unable to complete the self-report questionnaire and 1 questionnaire was not usable because of incomplete information. Regarding other informant reports, of the 251 participating ConHD patients, for 1 mentally retarded patient the questionnaire was not applicable, 2 patients did not have a suitable other informant because of language problems, 10 patients did not return the questionnaire and 4 questionnaires were not usable because of incomplete information.

The response rate for self-reports, corrected for mentally handicapped patients who were unable to complete the questionnaire, was 91.0%, resulting in usable questionnaires for 90.6% of the eligible patient sample. The response rate for other informant reports, corrected for mentally handicapped and persons with no other informant because of language problems, was 87.5%, resulting in usable questionnaires for 86.0% of the eligible patient sample.

Numbers of patients belonging to each diagnostic group were: closure of Atrial Septal Defect (ASD; n = 53), closure of Ventricular Septal Defect (VSD; n = 70), corrective surgery for Tetralogy of Fallot (ToF; n = 52), Mustard procedure for Transposition of the Great Arteries (TGA; n = 51) and surgery for Pulmonary Stenosis (PS; n = 25). The mean ages in these diagnostic groups were 29, 27, 27, 25 and 27 years respectively. Medical history and physical condition of these patients at the time of the first follow-up are described elsewhere (Meijboom et al., 1993; Meijboom et al., 1995; Meijboom et al., 1996; Meijboom et al., 1994).

The Dutch reference group consisted of 1441 young adults from the general population, with an age range of 20-32 years (mean age: 25 years) (Hofstra et al., 2000).

Instruments

At Time 1, emotional and behavioral problems in children, adolescents and young adults with ConHD were assessed with a parental questionnaire: the Child Behavior Checklist (CBCL; Achenbach, 1991a; Verhulst et al., 1996) and two self-reports for patients: the Youth Self-Report (YSR; Achenbach, 1991b; Verhulst et al., 1997) and the Young Adult Self-Report (YASR; Achenbach, 1990; Utens et al., 1998a; Utens et al., 1996; Utens et al., 1993).

At Time 2, emotional and behavioral problems were assessed with instruments parallel to those at Time 1: YASR (Achenbach, 1997) and a checklist to be completed by significant others, such as parents or a spouse: the Young Adult Behavior Checklist (YABCL; Achenbach, 1997).

Good reliability and validity of the YASR (Achenbach, 1997) have been confirmed for an earlier Dutch version of the YASR (Ferdinand et al., 1995). The YASR was originally developed for 18- to 30-year-olds and contains 110 problem items. The response format is 0 = not true, 1 = somewhat or sometimes true, and 2 = very true or often true. The YASR can be scored on 8 syndrome scales: Anxious/Depressed, Withdrawn, Somatic Complaints, Thought Problems, Attention Problems, Intrusive Behavior, Delinquent Behavior, and Aggressive Behavior, and two broad-band groupings of syndromes: Internalizing (consisting of the Anxious/Depressed and Withdrawn scales) and Externalizing (consisting of Intrusive Behavior, Delinquent Behavior and Aggressive Behavior scales). A Total Problems score is computed by summing the individual item scores.

The Young Adult Behavior Checklist (Achenbach, 1997) is the parallel version of the YASR, originally developed for 18- to 30-year-olds. It is to be completed by an informant who is familiar with the young adult, including a partner, parent or other close relative or friend. The YABCL contains 105 problem items and consists of the same scales as the YASR. Good reliability and validity was reported for the YABCL (Achenbach, 1997).

Assessment procedures

All patients were traced, approached uniformly and signed an informed consent before participating. During their visit to the Erasmus Medical Center Rotterdam, patients were interviewed and tested by a psychologist (EvR) and medically examined by a cardiologist. The YASR was filled in by the patient during the psychological examination. Some patients could not visit the hospital for practical or emotional reasons; they filled in the YASR at home and returned it by mail. The YABCL was filled in at home, by someone familiar to the patient and was returned by mail. For some patients, who were accompanied during their hospital visit by a familiar person (e.g. parent, spouse), the YABCL was completed in the waiting room.

Statistical methods

Differences in proportions scoring in the deviant range between patient sample versus reference group were analyzed by Binomial Testing. Differences in mean scale scores of the patient sample versus reference group were assessed with ANOVAs. Socioeconomic status (SES) was applied as a covariate if proven to have a main effect on the scale scores in the ANOVAs. In order to correct for chance findings, Bonferroni corrections were applied for the number of comparisons made. Odds Ratios and Pearson Correlation Coefficients were used to make longitudinal comparisons in the patient sample. Chi-squares were used to assess differences in sex, age, cardiac diagnosis and socioeconomic status between groups representing different trajectories across time.

Results

Proportions of problem adults

Table 3.1 shows the proportions of individuals in the ConHD patient sample and reference group, who scored in the deviant range of the YASR or the YABCL at Time 2. The 90th percentiles of the cumulative frequency distributions of the YASR and YABCL Total Problems scores obtained for the reference group were chosen as cut-offs, above which individuals scored in the deviant range (Achenbach, 1997). For both sexes, two age categories (20-27 and 28-32 years) were formed based on the median split in the patient sample. The percentages of patients who scored in the deviant range for the total sample and for females, aged 20 to 27 years, were significantly higher than those in the reference group, both for the YASR and YABCL. All the differences , reported in Table 3.1, remained significant after applying a Bonferroni correction for five comparisons.

Table 3.1 Percentages of the ConHD patient sample and reference group scoring in the YASR and YABCL deviant range at Time 2

	YASR			YABCL		
	% > Cut-off[a]		Binomial	% > Cut-off[a]		Binomial
Age categories	ConHD (n)	Reference (n)	Test *P*	ConHD (n)	Reference (n)	Test *P*
Males						
20 – 27[b]	16.2 (74)	9.9 (456)	ns	15.2 (66)	9.9 (414)	ns
28 – 32[b]	8.3 (60)	10.1 (207)	ns	13.3 (60)	9.4 (171)	ns
Females						
20 – 27[b]	27.8 (54)	9.7 (544)	< 0.01	30.9 (55)	9.3 (484)	<0.01
28 – 32[b]	13.0 (54)	10.3 (233)	ns	15.1 (53)	10.3 (185)	ns
Total sample	15.7 (242)	10.1 (1440)	< 0.01	18.4 (234)	10.3 (1254)	< 0.001

Note. Abbreviations used are: ConHD = Congenital Heart Disease, YASR = Young Adult Self-Report, YABCL = Young Adult Behavior Checklist.

[a] The 90th percentile of the cumulative frequency distribution of YABCL/YASR Total Problems scores obtained from the reference group.

[b] Age categories (20-27 versus 28-32 years) based on median splits of patient sample.

Mean problem scores

Table 3.2 shows the mean scale scores of the patient sample and reference group at Time 2. To test differences in mean problem scores, analyses of covariance (ANCOVAs) were computed in a group (ConHD versus reference) x sex (males versus females) x age (20-27 versus 28-32 years) factorial design. The influence of socioeconomic status was corrected for by using it as a covariate. In Table 3.2, the magnitude of significant ($p < 0.05$) group effects (ConHD versus reference) is indicated in terms of the percentage of variance accounted for. According to Cohen (1988), effects accounting for 1.0-5.9 % of variance are considered small, 5.9-13.8% medium and >13.8% large. Significant group effects found could thus be considered as small or <1%. All main effects for group, except for that on YASR Total Problems score, remained significant after applying a Bonferroni corrrection for 11 comparisons.

Significant group x age interaction effects were found for YASR Aggressive Behavior, YASR Externalizing, YABCL Thought Problems, YABCL Attention Problems, YABCL Intrusive and YABCL Total Problems score, and, though not significant after Bonferroni correction for 11 comparisons, for YASR Withdrawn, YASR Thought Problems, YASR Internalizing, YASR Total Problems score, YABCL Delinquent Behavior and YABCL Externalizing. All effects represented larger differences between the younger (20-27 years) and the older (28-32 years) individuals in the ConHD sample compared to the reference group, with the older ConHD patients scoring more favorably than the younger ConHD patients.

Table 3.2 Mean problem scores for YASR and YABCL scales for the ConHD patient sample and reference group at Time 2, and percentages of variance accounting for the difference between the ConHD and reference group in ANCOVAs

	YASR			YABCL		
Scales	Mean problem scores		Group[a] (%)	Mean problem scores		Group[a] (%)
	ConHD (n=242)	Reference (n=1438)		ConHD (n=234)	Reference (n=1253)	
Anxious/Depressed	5.1	5.1		4.3	3.4	1.0
Withdrawn	2.4	2.4		1.5	1.2	<1
Somatic Complaints	3.1	2.3	1.3	2.1	1.4	1.4
Thought Problems	0.2	0.2		0.6	0.4	<1
Attention Problems	2.3	2.3		4.1	2.6	3.0
Intrusive	2.2	2.2		2.1	1.4	2.2
Aggressive Behavior	2.5	2.3		3.1	2.1	1.2
Delinquent Behavior	1.0	0.9		0.6	0.7	
Internalizing	7.4	7.5		5.9	4.5	1.1
Externalizing	5.6	5.4		5.8	4.2	1.3
Total problems	29.6	27.5	<1[b]	23.2	16.2	2.5

Note. Abbreviations used are: YASR = Young Adult Self-Report, YABCL = Young Adult Behavior Checklist, ConHD = Congenital Heart Disease.

[a] Percentage of variance accounted for by significant group effect (ConHD versus Reference).

[b] Not significant after Bonferroni correction for 11 comparisons.

Significant (though not after Bonferroni correction for 11 comparisons) group x sex interaction effects were found for YABCL Anxious/Depressed, YABCL Attention Problems, YABCL Intrusive, YABCL Aggressive Behavior, YABCL Externalizing and YABCL Total Problems score. The effects on YABCL Anxious/Depressed, YABCL Aggressive Behavior and YABCL Total Problems score represented larger differences between males and females in the ConHD sample compared to the reference group, with males scoring more favorably than females in both the patient sample and reference group. On YABCL Attentions Problems, YABCL Intrusive and YABCL Externalizing, ConHD males scored more favorably than ConHD females, whereas in the reference group females scored more favorably than males.

Significant (though not after Bonferroni correction for 11 comparisons) group x age x sex interaction effects were found for YASR Intrusive, YABCL Thought Problems and YABCL Total Problems score. These effects indicated larger differences between younger (20-27 years) and older (28-32 years) individuals among the ConHD females, compared to the females in the reference group, with the older ConHD females scoring more favorably than the younger ConHD females.

Different informants

On the YASR, differences between the ConHD patient sample and reference group are limited to the specific problem area of Somatic Complaints. On the YABCL, the ConHD patient sample scored less favorably than the reference group on almost all problem areas, except for Delinquent Behavior. Since YASR and YABCL were filled in by different informants, informant effect was examined by a repeated measures design for YASR and YABCL scores at Time 2. A significant informant x group interaction effect was found ($p < 0.05$), indicating a differential informant effect. That is, greater difference between the ConHD patient sample versus the reference group was found for the YABCL than for the YASR. However, the YABCL for the ConHD patient sample was completed by significant others (mostly partners) for 61.5% and by parents for 38.5%, whereas the YABCL in the reference group was completed by significant others for 3.3% and by parents for 96.7%. Therefore, within the ConHD patient sample, an ANOVA with an informant (parent versus significant other) x sex (males versus females) x age (20-27 years versus 28-32 years) factorial design was computed on YABCL Total Problems score. No significant main- or interaction effect for informant was found, indicating that there was no significant difference in scoring by parents versus significant others.

Diagnostic groups

Table 3.3 shows the mean Total Problems scores and mean scale scores of the different cardiac diagnostic groups within the patient sample at Time 2. To test differences in mean problem scores, ANOVAs were computed for a cardiac diagnosis (5 diagnoses) x sex (males versus females) x age (20-27 years versus 28-32 years) factorial design. One main effect for diagnosis was found for YABCL Attention Problems ($p < 0.05$). Post hoc tests of least squared differences revealed that patients with VSD and ToF scored significantly higher on Attention Problems than patients with ASD (respectively $p < 0.001$, and $p < 0.01$). Patients with VSD scored significantly higher on Attention Problems than patients with PS ($p < 0.05$). After applying Bonferroni correction for 11 comparisons, no main or interaction effect remained significant.

Table 3.3 Mean problem scores of YASR and YABCL for different cardiac diagnostic groups at Time 2

	YASR (n=242)						YABCL (n=234)					
Scales	ASD (n=53)	VSD (n=66)	ToF (n=49)	TGA (n=51)	PS (n=23)	Main effects	ASD (n=50)	VSD (n=65)	ToF (n=50)	TGA (n=45)	PS (n=24)	Main effects
Anxious/Depressed	5.1	5.2	4.1	6.0	4.6	ns	4.1	4.5	3.9	4.8	4.5	ns
Withdrawn	2.1	2.3	2.3	3.0	2.1	ns	1.6	1.5	1.4	1.7	1.6	ns
Somatic Complaints	3.1	3.0	2.7	3.4	3.1	ns	2.1	1.9	2.3	2.2	2.1	ns
Thought Problems	0.1	0.3	0.2	0.3	0.1	ns	0.5	0.7	0.8	0.6	0.4	ns
Attention Problems	2.1	2.3	2.1	2.7	2.2	ns	2.8	5.2	4.5	4.0	3.3	$< 0.05^a$
Intrusive	1.8	2.5	1.8	2.5	2.0	ns	1.7	2.6	2.2	2.1	1.8	ns
Aggressive Behavior	2.0	2.5	2.2	3.4	1.9	ns	3.2	3.8	2.7	3.0	2.3	ns
Delinquent Behavior	0.6	1.2	0.9	1.5	0.7	ns	0.4	0.8	0.3	1.0	0.2	ns
Internalizing	7.2	7.5	6.3	9.0	6.7	ns	5.7	6.0	5.3	6.4	6.0	ns
Externalizing	4.4	6.2	5.0	7.3	4.5	ns	5.3	7.2	5.2	6.1	4.3	ns
Total Problems	27.1	30.5	27.0	34.9	26.7	ns	21.4	26.1	22.7	23.3	19.9	ns

Note. Abbreviations used are: YASR = Young Adult Self-Report, YABCL = Young Adult Behavior Checklist, ASD = Atrial Septal Defect, VSD = Ventricular Septal Defect, ToF = Tetralogy of Fallot, TGA = Transposition of the Great Arteries, PS = Pulmonary Stenosis.

[a] Not significant after Bonferroni correction for 11 comparisons.

Longitudinal course

Longitudinal comparisons were made to examine the course of psychopathology in ConHD patients across the 10 year follow-up interval. Only comparisons were made with the same type of informant (self or other) at Time 1 and Time 2 (significant others: CBCL-YABCL, self-reports: YSR-YASR and YASR-YASR). At Time 1, not all subjects had reached adulthood. Then parallel instruments were used to measure psychopathology in childhood (CBCL), adolescence (YSR) and adulthood (YASR). For 83 patients with an overlapping age range (10-15 years) at Time 1, both CBCL- and YSR-data were available. These patients are included in CBCL-YABCL as well as YSR-YASR comparisons. According to Cohen's (1988) criteria medium correlations were found for the CBCL-YABCL (r = 0.34) and YSR-YASR comparisons (r = 0.46) and a large correlation was found for the YASR-YASR comparison (r = 0.64). All of these positive correlations were significant at a 0.01 level.

For a total of 220 patients, data were available at both Time 1 and Time 2. Table 3.4 shows the odds ratios for these three longitudinal comparisons of scoring in the deviant range on Total Problems score for both sexes as well as for the total sample. The predictive value of scoring in the deviant range at Time 1 for scoring in the deviant range at Time 2 is highest for the YASR-YASR comparison and higher for males than for females. That is, for older and male patients, the probability that psychopathological symptoms will remain persistent in the future is higher than for younger and female patients respectively.

Table 3.4 Odds Ratios for Time 1 and Time 2 Total Problems scores in the deviant range

Time 1 – Time 2	OR (95% CI)		
	Males	Females	Total
CBCL – YABCL (n=93)	6.2 (1.4-27.9)	1.6 (0.4-6.9)	3.1 (1.1-8.8)
YSR – YASR (n=127)	5.1 (1.4-18.8)	2.0 (0.6-7.3)	3.4 (1.4-8.5)
YASR – YASR (n=93)	- [a]	3.9 (0.3-51.2)	21.6 (3.8-124.1)

Note. Abbreviations used are: OR = Odds Ratio, CBCL = Child Behavior Checklist, YABCL = Young Adult Behavior Checklist, YSR = Youth Self-Report, YASR = Young Adult Self-Report.
[a] Odds Ratio can not be computed due to an empty cell.

To take a closer look at the development of psychopathology in the ConHD patient sample, different groups representing different trajectories were formed, based on scoring in or out the deviant range at Time 1 and Time 2 (see Table 3.5). Cut-offs were computed for males and females separately. “Normals” did not score in the deviant range at neither Time 1 or Time 2. “Increasers” did not score in the deviant range at Time 1, but did so at Time 2. “Decreasers” did score in the deviant range at Time 1, but not at Time 2. “Persisters” scored in the deviant range at both Time 1 and Time 2.

Chi-Squares were computed for the trajectory groups in the YSR-YASR and YASR-YASR comparisons (N = 220) for which patients themselves were informant at both Time 1 and Time 2. Sex (males versus females), age (20-27 versus 28-32 years), cardiac diagnosis (5 diagnoses) and Socioeconomic status (low, middle, high) were applied as grouping variables. A significant difference in proportions of trajectory groups was found for age ($\chi^2 = 16.15$; *df* = 3; $p < 0.01$). The 20-27-year-olds belonged to the "Increasers", "Decreasers" and "Persisters" more often than the 28-32-year-olds; the 28-32-year-olds belonged to the "Normals" more often than the 20-27-year-olds. No significant differences were found for sex, cardiac diagnosis and social economic status. Chi-squares for stable trajectories ("Normals" and "Persisters") versus trajectories reflecting change over time ("Increasers" and "Decreasers") revealed significant differences for sex ($\chi^2 = 4.72$; *df* = 1; $p < 0.05$) and age ($\chi^2 = 11.77$; *df* = 1; $p < 0.01$). Males and the 28-32-year-olds belonged to the stable trajectories more often than females and 20-27-year-olds respectively. No significant differences were found for cardiac diagnosis and social economic status. Table 3.5 shows the proportions of the ConHD patient sample in different trajectory groups, specified by sex and age.

Table 3.5 Percentages of the ConHD patient sample in different trajectory groups[a]

	% Normals (n)	% Increasers (n)	% Decreasers (n)	% Persisters (n)	% Total (n)
Males					
20 – 27 years[b]	67.2 (43)	10.9 (7)	12.5 (8)	9.4 (6)	100 (64)
28 – 32 years[b]	91.4 (53)	1.7 (1)	1.7 (1)	5.2 (3)	100 (58)
total	78.7 (96)	6.6 (8)	7.4 (9)	7.4 (9)	100 (122)
Females					
20 – 27 years[b]	54.3 (25)	15.2 (7)	19.6 (9)	10.9 (5)	100 (46)
28 – 32 years[b]	78.8 (41)	5.8 (3)	11.5 (6)	3.8 (2)	100 (52)
total	67.3 (66)	10.2 (10)	15.3 (15)	7.1 (7)	100 (98)
Total sample	73.6 (162)	8.2 (18)	10.9 (24)	7.3 (16)	100 (220)

Note. Abbreviation used is: ConHD = Congenital Heart Disease.

[a]Only YSR-YASR and YASR-YASR comparisons included.

[b]Age categories (20-27 versus 28-32 years) based on median splits of patient sample.

Discussion

Level of psychopathology in the patient sample compared to the reference group

Both patients' (YASR) and significant others' (YABCL) reports showed significant greater proportions of individuals who scored in the deviant range in the total ConHD patient sample versus the reference group. Significantly higher mean problem scores for the ConHD patient sample were found on the YABCL. According to the self-reports (YASR), significantly higher mean problem scores for the ConHD patient sample were found for Somatic Complaints only. At Time 1, Utens et al. (1993) found greater proportions falling in the psychopathological range among ConHD children and adolescents; for ConHD adults they found such elevated proportions to a much lesser extent (Utens et al., 1998a). In contrast to our present findings, they found a greater proportion of young adult male patients who scored in the deviant range, instead of female patients. Further results by Utens et al. (1998a) resemble those of the present study. The only significant difference found on Somatic Complaints might be explained by actual symptoms in the ConHD patient sample, such as heart pounding or palpitation.

Effect of sex, age and cardiac diagnosis on present level of psychopathology

Proportions of individuals in the ConHD patient sample who scored in the deviant range were especially high among 20-27-year-old females. Although not always significant after Bonferroni correction for 11 comparisons, the interaction effects found including the factor group were all in the same direction. Compared to the reference group, greater differences were found between males and females in the ConHD group, with males scoring more favorably than females in both the patient sample and reference group. For some scales ConHD females scored less favorably than ConHD males, while in the reference group the reversed situation was observed. Also, compared to the reference group, greater differences between patients in early and later adulthood were observed, with patients from later adulthood scoring more favorably than those from early adulthood. For a few scales, this difference was found especially among ConHD females. Overall, especially young adult female ConHD patients seem to be at risk for developing psychopathological symptoms. A possible explanation might be that young ConHD women experience specific, disease-related uncertainties (Gantt, 1992; Wright et al., 1985). The issue of sexual relationships and offspring might raise uncertainties regarding acceptation and abilities of pregnancy and delivery. Presumably, these uncertainties might strike young ConHD women in particular since these issues involve their own bodily functioning. This explanation is supported by theoretical model by Cyranowski et al. (2000) who state that females have a preference for close emotional communication, intimacy and responsiveness within interpersonal relationships and develop a sense of self in relation to others, whereas males are more focussed on personal autonomy. According to Cyranowski et al. (2000) this difference between sexes explains why young females are more likely than males to become depressed when faced with negative life events, especially when these life events involve interpersonal consequences. Therefore, negative consequences of the ConHD on life events which are prominent in early adulthood, such as relations and offspring, are likely to strike females harder than males. These possible causal factors for psychopathology in young ConHD females might recede more to the background when a stable relationship and family life are established, which might explain the higher decline of psychopathological symptoms from early- to later adulthood in ConHD patients compared to the reference group.

Only one significant main effect for cardiac diagnosis was found, indicating more attention problems in VSD and ToF versus ASD and also in VSD versus PS. After Bonferroni correction, this effect did not remain significant. This confirms earlier findings (Brandhagen et al., 1991; Utens et al., 1998a; Utens et al., 1993).

Effect of different informants

Other informants seem to evaluate psychopathological symptoms in adult ConHD patients as more deviant from the norm than patients themselves. This finding seems to confirm previously suggested denial and overcompensation in ConHD patients (Utens et al., 1994), which might lead to a tendency to underestimate their emotional and behavioral problems. A differential informant effect was found, indicating a greater difference between self and other informant report in the ConHD patient sample compared to the reference group, with others indicating more problems than patients themselves. This effect could not be explained by the different types of informants, since no significant effect for informant (parent versus significant other) was found within the ConHD patient sample.

Development of psychopathology in the patient sample

Since at Time 1 psychopathology was measured with similar instruments as at Time 2, this study offered the unique possibility to make longitudinal comparisons of prevalence of psychopathology in ConHD patients over a period of 10 years. In the present study, the predictive value of psychopathological symptoms for future behavioral or emotional problems and matching Pearson correlation coefficient were higher for the adolescence–adulthood comparisons (YSR-YASR) than for the childhood–adulthood comparisons (CBCL-YABCL), and highest for the early-later adulthood comparisons (YASR-YASR). Thus, the predictive value of psychopathological symptoms for future behavior or emotional problems increases with age.

Effect of sex, age, cardiac diagnosis and social economic status on the development of psychopathology

In analyses based on trajectory groups, the relatively older patients more often belonged to the "normals" and more often followed stable trajectories in contrast to the younger patients. Male patients followed stable trajectories more often than female patients, whereas female patients showed more change across time in their trajectories. Cardiac diagnosis and social economic status did not seem to influence the trajectories of psychopathology. Overall, psychopathology seems to decrease as patients grow older. This trend is also seen in the general population (Hofstra et al., 2000). However, the decline of psychopathology in the ConHD patient sample was revealed when comparing to a reference groups and can therefore be characterized as stronger and more prominently present than in the general population.

Limitations

It should be noted that the present sample of ConHD patients contains a selection of 5 large diagnostic groups within specific age ranges and is therefore not completely representative for all ConHD anomalies. Relative small sizes of the 5 separate diagnostic groups might hamper revealing differences between these groups.

Because different, though similar instruments were used at Time 1 and Time 2, repeated measures analyses were not possible for all patients. Analyses based on categorization in trajectories of psychopathology provide an rough image of the development of psychopathology in adult ConHD patients. Influence of specific medical based predictors (medical data not yet systematically available) should be described in future research.

Not all effects described were significant after Bonferroni correction. Effects found represented differences in the same direction and were therefore not considered as coincidental products of multiple testing, but as real trends in the ConHD patient sample.

Implications

An elevated level of psychopathology is seen in the total sample of ConHD patients, which can be mainly attributed to the young adult ConHD patients. Among older ConHD patients, psychopathological symptoms are less prominently present, but show a higher predictive value for future problems. On the one side this indicates that behavioral and emotional problems in ConHD patients seem to diminish when patients grow older, probably because specific uncertainties recede to the background in adulthood. On the other side, these findings imply that assessment and treatment of psychopathology at young age is highly desirable in

ConHD patients. Not only do psychopathological symptoms seem less persistent and thus better treatable at a young age, dealing with disease-related uncertainties might make the ConHD patients' struggle in early adulthood less hard and therefore improve their quality of life. Providing information could help ConHD patients to deal with uncertainties they are facing concerning their physical health, childbearing abilities, etc. Special attention should be given to specific fears and uncertainties young female ConHD patients are facing. In order to pay attention to these problems adequately, more information is needed about how young ConHD women judge and experience their abilities.

When assessing or treating psychopathology in ConHD patients, awareness for denial mechanisms or overcompensation in ConHD patients should be taken into account. This study revealed that parents and partners of ConHD patients report more behavioral and emotional problems compared to the reference group whereas ConHD patients themselves hardly did so. This finding is an important indication for underestimation of behavioral and emotional problems in ConHD adults.

4

Coping styles and social support in a cohort of adults with congenital heart disease

E.H.M. van Rijen

E.M.W.J. Utens

J.W. Roos-Hesselink

F.J. Meijboom

R.T. van Domburg

J.R.T.C. Roelandt

A.J.J.C. Bogers

F.C. Verhulst

Chapter 4

Coping styles and social support in a cohort of adults with congenital heart disease

Abstract

Objective: *To determine coping styles and social support in a cohort of adults with congenital heart disease (ConHD).* ***Methods:*** *342 ConHD adults (20-46 years), who received their first heart surgery in childhood, filled in questionnaires concerning coping styles (Utrecht Coping List) and social support (Social Support List).* ***Results:*** *Compared to the reference group, the total ConHD patient sample reported to receive less social support, but also experienced less discrepancies between desired and received social support. ConHD females reported to seek and receive more social support than ConHD males. Considering the relationship with social support however, ConHD females reported less favourable coping styles compared to reference females, such as less active problem solving. In contrast, ConHD males had coping styles more favourably related to social support at their disposal, compared to reference males.* ***Conclusion:*** *The less active problem solving in ConHD females needs special attention.*

Introduction

Over the last decades, major advances in surgical intervention for congenital heart disease (ConHD), have lead to increasing survival rates for ConHD patients. Therefore, an increasing patient population of adults with ConHD is emerging today. Since these ConHD patients belong to the first generation of patients who has reached adulthood, they are also the first patients to face specific problems that ConHD in adulthood involves. These problems concern a wide range of aspects of adult life, such as employability and insurability (Allen et al., 1992; Celermajer & Deanfield, 1993; Gersony et al., 1993; Hart & Garson, 1993; Hellstedt, 1994; Mahoney et al., 1991; McGrath & Truesdell, 1994) as well as sexuality and childbearing (Gantt, 1992; Mahoney et al., 1991; Patton et al., 1990; Somerville, 1998; Tong & Sparacino, 1994). Also, emotional and social problems in ConHD adults have been reported (Brandhagen et al., 1991; Shampaine et al., 1990; van Rijen et al., submitted b).

The way parents of ConHD children deal with their child's illness is well documented (Davis et al., 1998; Gardner et al., 1996; Garson et al., 1978; Stinson & McKeever, 1995; Utens et al., 2000a; Utens et al., 2002). Surprisingly, very little is known about how ConHD patients themselves cope with problems they come across as they grow older. Overcompensation and denial of problems associated with the ConHD have been suggested as coping strategies in adult ConHD patients (Ternestedt et al., 2001; Utens et al., 1996; Utens et al., 1994). So far however, coping styles in ConHD adults have not been examined explicitly. More knowledge about coping styles in ConHD adults might be helpful in understanding how they experience potential uncertainties and disabilities their disease brings along, since often, psychological and social consequences do not seem to be related to the severity of the cardiac diagnosis (Brandhagen et al., 1991; Utens et al., 1996; Utens et al., 1994; van Rijen et al., submitted b).

Research into the role of the social environment in the ConHD patients' functioning has indicated parental anxiety, overprotection and pampering (Gupta et al., 1998; Kokkonen & Paavilainen, 1992; Linde et al., 1966; Utens et al., 1994) as risk factors for developing long term psychosocial problems. However, the social environment of ConHD patients at adult age, in which the role of parents is generally diminished, has not yet been explored. More knowledge about how ConHD adults feel they are dealt with by their social environment and how it affects them is required to understand their feelings and attitudes.

Since coping styles in ConHD adults are likely to be connected to the social support in their present life, these aspects will be closely examined in this study. The aims of the present study are:

1. To compare current coping styles and social support of adult patients (age range 20-46 years), operated for ConHD at a young age, with those of normative groups of similar age.
2. To identify the role of sex, age and cardiac diagnosis in coping styles and social support in adult patients operated for ConHD at a young age.
3. To examine the relationship between coping styles and social support in adult patients operated for ConHD at a young age.

Method

Inclusion criteria

During the first follow-up study in this cohort of patients, which took place in 1989-1991, all consecutive patients who underwent their first open heart surgery for congenital heart disease between 1968 and 1980 in the Erasmus MC, and who were younger than 15 years at the time of surgery, were eligible. This population is described in detail elsewhere (Utens et al., 1998a; Utens et al., 1994; Utens et al., 1993).

The target population of the second follow-up (2000-2001) consisted of the 498 patients of the first follow-up. From the 498 patients we excluded 61 patients who belonged to a miscellaneous diagnostic group, consisting of small numbers of patients with a variety of congenital heart defects (aortic stenosis, discrete subaortic stenosis, primum type atrial septal defect, complete atrioventricular septal defect, pulmonary atresia, tricuspid atresia, total abnormal pulmonary venous drainage, truncus arteriosus, and miscellaneous diagnoses). Furthermore we excluded 11 deceased patients, 26 untraceable patients and 1 patient who had undergone heart transplantation.

Patient sample

Of the 399 eligible patients, 37 refused to participate. Among the remaining 362 patients, 17 mentally retarded patients and 1 patient with a language problem were not able to respond to the questionnaires; 2 patients who preferred to fill in the questionnaires at home, did not return these completely. The overall response rate, corrected for deceased persons and persons lost to follow-up was 90.7%, resulting in usable questionnaires for this particular study for 85.7% of the eligible patient sample (n = 342, 185 males, 157 females, mean age 30.3 years, age range 20-46 years).

Numbers of patients belonging to different diagnostic groups were: closure of Atrial Septal Defect (ASD, n = 92), closure of Ventricular Septal Defect (VSD, n = 86), corrective surgery for Tetralogy of Fallot (ToF, n = 72), Mustard procedure for Transposition of the Great Arteries (TGA, n = 55) and surgery for Pulmonary Stenosis (PS, n = 37). The mean ages at the time of study in these diagnostic groups were 32.9 (sd 4.6, range 21.0-43.9), 29.7 (sd 5.6, range 21.2-43.6), 30.2 (sd 5.6, range 20.9-45.2), 26.0 (sd 4.0, range 20.5-37.1), 31.5 (sd 5.7, range 23.1-46.2) years respectively. Medical history and physical condition of these patients at the time of the first and second follow-up are described elsewhere (Meijboom et al., 1993; Meijboom et al., 1995; Meijboom et al., 1996; Meijboom et al., 1994; Roos-Hesselink et al., 2003).

Reference group

Normative data were derived from the published manuals of the questionnaires used. The male and female reference groups for the Utrecht Coping List (Schreurs et al., 1993) consisted of employers of the Dutch railways and hospital nurses respectively, both combined with a random sample of the general population. Age ranges of the male (n = 599) and female (n = 542) reference groups were respectively 19-45 years and 18-45 years. The reference group for the Social Support List (van Sonderen, 1993) was derived from different studies among non-patient groups. The total reference group consisted of 786 persons (mean age 34.3 years) of which 307 were male and 479 female. Data were collected for 734 persons.

Instruments

Utrecht Coping List. The Utrecht Coping List (UCL) is a reliable and standardised self-report of styles of coping (Schreurs et al., 1993). The respondent is asked to indicate how often he/she reacts to problems in a certain manner. The response categories are: seldom or never; sometimes; often; very often. Styles of coping are measured on 7 scales, namely: Active Solving of Problems (approaching problems in a purposeful and confident manner), Palliative Reaction (seeking diversion in an healthy or unhealthy manner), Avoiding/Waiting (avoiding difficult situations or waiting to see what happens), Seeking Social Support (sharing feelings), Passive Patterns of Reaction (being absorbed by problems and showing hopelessness), Expression of Emotions (showing aggression and annoyance) and Reassuring Thoughts (using positive cognitions).

Social Support List. The Social Support List (SSL; van Sonderen, 1993) measures the interactions and discrepancies that people experience in receiving social support from their direct environment. Interactions represent the subjective experience of frequency of receiving social support. The response categories are: seldom or never; sometimes; frequently; very often. Discrepancies represent the subjective experience of mismatches between the received and desired frequency of receiving social support. The response categories are: I miss it; I don’t really miss it, but I prefer more; exactly the right amount; it happens too often. The latter two response categories were combined into one score. Interactions as well as Discrepancies are measured on 6 dimensions of social support, namely: Daily-oriented Emotional Support (affection or support in daily situations), Problem-oriented Emotional Support (emotional support in difficult situations), Esteem (appreciation shown by e.g. compliments or being asked for advice), Instrumental Support (practical help or support in a material way or by services), Social Companionship (social contacts or involvement in social activities), and Informative Support (information about one’s behaviour). For a separate additional scale of Negative Reactions, only interactions were measured. A total score was calculated for interactions and discrepancies of social support separately. For some dimensions, shorter terms were used in the tables of this study. Overall, satisfying reliability was reported for the Social Support List (van Sonderen, 1993). Reliability was poorer for Instrumental and Informative Support.

Assessment procedures

All patients were traced, approached uniformly and signed an informed consent before participating in the study. During their visit to the Erasmus MC, patients were interviewed and tested by a psychologist (EvR) and medically examined by a cardiologist. Some patients could not visit the hospital for practical or emotional reasons; they filled in the questionnaires at home and returned it by mail.

Statistical analyses

One sample t-tests, based on 95% confidence intervals, were used to test differences between the patient sample and reference groups on the UCL and SSL. To identify the role of sex (male, female), age (20-29 years, 30-46 years) and cardiac diagnosis (ASD, VSD, ToF, TGA and PS), univariate analyses of covariance (ANCOVAs), based on 95% confidence intervals, were performed on all SSL- and UCL-scales. Because the variables relationship (relationship, no relationship) and socioeconomic status (SES; low, middle, high) both showed significant main effects in ANOVAs of about half of UCL- and SSL-scales, they were applied as covariates. In order to correct for multiple comparisons in analyses mentioned above, Bonferroni corrections were applied. Pearson correlations between UCL- and SSL-scales were calculated to examine the relationship between coping styles and social support in the ConHD patient sample.

Results

Coping styles

Table 4.1 shows the mean scores on the Utrecht Coping list for the ConHD patient sample and reference group. ConHD males obtained significantly higher scores on Seeking Social Support and lower scores on Passive Patterns of Reaction and Expression of Emotions, compared to the reference group. ConHD females scored significantly lower on Active Solving of Problems compared to the reference group. For the total patient sample, less active problem solving was found. After Bonferroni correction, no further differences in coping styles between ConHD patient sample versus reference group were found.

Table 4.1 Mean scores of the ConHD patient sample and reference group on the Utrecht Coping List

	Males			Females			Total		
Scales	ConHD (N=185)	Ref (N=599)	*P*	ConHD (N=157)	Ref (N=542)	*P*	ConHD (N=342)	Ref (N=1141)	*P*
Active Problem Solving	18.6	18.3	ns	17.2	18.8	< 0.001	18.0	18.5	0.004
Palliative Reaction	15.6	15.3	ns	16.8	16.8	ns	16.2	16.0	ns
Avoiding/Waiting	14.7	14.7	ns	15.0	14.6	0.029[a]	14.8	14.7	ns
Seeking Social Support	12.5	11.3	0.001	14.2	14.7	0.031[a]	13.3	12.9	0.019[a]
Passive Reaction Pattern	9.8	10.6	0.001	10.7	10.3	0.037[a]	10.2	10.5	0.039[a]
Expression of Emotions	6.0	6.3	0.003	6.4	6.4	ns	6.2	6.3	ns
Reassuring Thoughts	11.2	11.6	0.021[a]	11.7	11.6	ns	11.5	11.6	ns

Note. A high score indicates high report of the relevant coping style.
[a] Not significant after Bonferroni correction for 7 comparisons.

Social support

Table 4.2 shows the mean scores on the Social Support List for the ConHD patient sample and reference group. The ConHD patient sample reported significantly less interactions concerning the scales Problem-oriented Emotional Support, Instrumental Support, Informative Support, Total Interactions and Negative Reactions, compared to the reference group. Also, significantly less discrepancies were reported by the ConHD patient sample on the scales Problem-oriented Emotional Support, Esteem, Informative Support and Total Discrepancies, compared to the reference group. Further analysis showed that both ConHD males and females experience significantly less discrepancies compared to reference males and females respectively (p<0.001 and p=0.002 respectively, data not shown).

Table 4.2 Mean scores of the ConHD patient sample and reference group on the Social Support List

	Interactions			Discrepancies		
Scales	ConHD (N=342)	Reference (N=734)	*P*	ConHD (N=342)	Reference (N=734)	*P*
Daily Emotional	10.3	10.6	0.022[a]	5.3	5.5	0.013[a]
Problem Emotional	16.8	17.6	< 0.001	9.8	10.4	< 0.001
Esteem	14.7	14.9	ns	7.5	7.8	< 0.001
Instrumental Support	13.2	14.5	< 0.001	8.0	8.2	0.025[a]
Social Companionship	13.4	13.4	ns	6.6	6.8	0.017[a]
Informative	8.1	8.6	< 0.001	5.1	5.6	< 0.001
Total	76.5	79.6	< 0.001	42.2	44.3	< 0.001
Negative Reactions	9.9	10.7	< 0.001			

Note. A high score indicates high report of the relevant interaction or discrepancy in social support.
[a] Not significant after Bonferroni correction for 15 comparisons.

Effects of sex, age and cardiac diagnosis

After applying Bonferroni correction for 7 comparisons, significant main effects for sex within the patient sample remained on the UCL-scales Active Solving of Problems, Palliative Reaction and Seeking Social Support. ConHD males reported higher Active Solving of

Problems than ConHD females (*p*<0.001). ConHD females scored higher on Palliative Reaction (*p*=0.007) and Seeking Social Support (*p*<0.001) than ConHD males. No significant main effect for age or cardiac diagnosis was found. A significant diagnosis x age interaction effect was found on Passive Patterns of Reaction (*p*=0.006). In the ToF- and PS-groups, the older patients (30-46 years) obtained higher scores on Passive Patterns of Reaction than the younger patients (20-29 years), whereas in the ASD-, VSD- and TGA-groups, the younger patients scored higher than the older patients.

After applying Bonferroni correction for 11 comparisons, significant main effects for sex within the patient sample were found on the SSL-scales of interactions Daily-oriented Emotional Support (*p*<0.001), Problem-oriented Emotional Support (*p*<0.001), Social Companionship (*p*=0.002) and Total Interactions (*p*=0.001), all indicating higher scores for the ConHD females compared to the ConHD males. No significant main effect for age or cardiac diagnosis was found. A sex x age interaction effect was found for discrepancies in Instrumental Support (*p*=0.002). Among ConHD males, older patients (30-46 years) reported more discrepancies than younger patients (20-29 years), whereas the older female ConHD patients reported less discrepancies than the younger female ConHD patients.

Relation between coping styles and social support

In order to examine the relationship between coping styles and subjective experiences of social support in ConHD patients, Pearson correlations were calculated between the UCL-scales and SSL-scales Total Interactions, Negative Reactions and Total discrepancies. Except for 4 correlations, all correlations shown in Table 4.3 were significant at a significance level of 1% or 5%. All significant correlations were positive, except for the correlation between Total Discrepancies and Active Problem Solving, which was found to be negative. According to Cohen (1988), correlations of 0.10 to 0.29 are considered small, correlations of 0.30 to 0.49 are considered medium and correlations above .50 are considered large. Medium positive correlations were found between the Total Interactions and the coping styles Palliative Reaction, Seeking Social Support and Reassuring Thoughts. A medium positive correlation was found between Negative Reactions and the coping style Passive Patterns of Reaction. A medium positive correlation was found between Total Discrepancies and the coping style Passive Patterns of Reaction. Further correlation analyses between and all separate interaction- and discrepancy-scales and the coping styles revealed positive correlations ≥ 0.40 between interactions of Esteem and the coping style Active Solving of Problems, between interactions of Daily-oriented Emotional Support and the coping style Seeking Social Support and between discrepancies of Daily-oriented Emotional Support, Problems-oriented Emotional Support and Social Companionship and the coping style Passive Patterns of Reaction.

Table 4.3 Pearson correlations between Utrecht Coping List and Social Support List

	SSL Scales		
UCL Scales	Total Interactions	Negative Reactions	Total Discrepancies
Active Problem Solving	0.26	0.03[b]	-0.18
Palliative Reaction	0.36	0.28	0.14
Avoiding/Waiting	0.14[a]	0.27	0.11[a]
Seeking Social Support	0.47	0.05[b]	-0.07[b]
Passive Reaction Pattern	0.14	0.45	0.44
Expression of Emotions	0.21	0.28	0.15
Reassuring Thoughts	0.35	0.14[a]	-0.02[b]

Note. All correlations were significant at $p<0.01$ except those marked with [a] or [b].
[a] Correlation significant at $p<0.05$.
[b] Correlation not significant.

Discussion

Coping Styles

The present study showed significantly lower Active Problem Solving for the total patient sample compared to the reference group. This result can be attributed to the ConHD females, since they showed a lower mean score on Active Problem Solving, compared to the reference group. ConHD females further reported to avoid problems or wait more often, seek social support less often and have passive patterns of reaction more often than the reference group. Although the latter three differences did not remain significant after Bonferroni correction, they were all disadvantageous and can therefore be considered as showing a trend in negative direction for ConHD females. In contrast, ConHD males showed coping styles which can be considered as favourable compared to the reference group, since they reported to seek social support (share feelings) more often, have less passive patterns of reactions and express emotions of anger and annoyance less often than the reference group.

Remarkably, similar results were found for styles of coping in parents of children awaiting elective cardiac surgery (Utens et al., 2000a). Mainly the mothers showed less favourable coping styles, compared to a reference group. Mothers of children with congenital heart disease awaiting surgery had a less active style of solving problems, and a more passive reaction pattern, than did reference females. Thus, the present results seem in line with a previous study, although different target samples concerning different types of ConHD were used. Possibly, mothers of ConHD patients, being the female role model, might transfer a less active and more passive coping style to their daughters. Previously, the negative influence of parental anxiety and coping on the development of children with ConHD has been mentioned (Masi & Brovedani, 1999; Utens, 1992). Unfortunately, no longitudinal study to examine the influence of parental coping styles into ConHD patients' functioning in adulthood has yet been performed. Another explanation for our findings could be that females deal with stressful life events using less adequate coping styles than males. This is in line with the earlier suggested explanation for increased levels of psychopathology found among the young adult ConHD females of this same cohort (van Rijen et al., submitted b). This suggested explanation is based on the theoretical model of Cyranowski et al. (2000), which states that females tend to display a strong preference for close emotional communication, intimacy, and responsiveness within interpersonal relationships and are therefore more vulnerable to become depressed when faced with negative life events, especially those with interpersonal consequences. Disease-specific uncertainties concerning life events which are prominent in adulthood, such as relations and offspring might therefore strike ConHD females harder than ConHD males.

Social support

Remarkably, the ConHD patient sample overall reported significant less interactions of social support, but also significant less experience of discrepancies between desired and received social support, compared to the reference group. The ConHD patients reported less interactions concerning problem-oriented emotional support, practical help, feedback on one's behaviour and negative reactions compared to the reference group. Combined with the significant less discrepancies they reported on emotional support in difficult situations, appreciation by others and feedback on one's behaviour, these results might indicate that the ConHD patients of the present study feel rather independent. Especially for emotional support when facing problems or for feedback on one's behaviour they don't seem to need too much support of their environment, or at least give the impression not to do so. Possibly, this could be a counteraction or overcompensation following the overprotection and pampering by the social environment, which have been suggested to occur in ConHD patients (Gupta et al., 1998; Kokkonen & Paavilainen, 1992; Linde et al., 1966; Utens et al., 1994). Carefulness must be taken into account in the interpretation of these results, since denial mechanisms concerning the negative consequences of the ConHD have been previously reported in ConHD patients (Utens et al., 1994; van Rijen et al., submitted b). Concerning appreciation by others and social companionship, no differences in interactions were found between the patient sample and reference group.

Effects of sex, age and cardiac diagnosis

The main effects of sex on coping styles in the patient sample seem to correspond with differences between males and females in the reference group, except for Active Problem Solving. On this scale, reference females scored higher than reference males, whereas ConHD females showed significant lower active problem solving compared to ConHD males. The reason why, in contrast to other diagnostic categories, in the ToF- and PS-groups, older patients showed more passive patterns of reaction than younger patients, is unclear.

The greater social support received in ConHD females compared to ConHD males appears in emotional support in daily of problem situations and social companionship. The increased experienced discrepancy between desired and received instrumental support in older ConHD males might indicate a prolonged dependency on practical help, which young adults are more likely to receive than older adults.

Relation between coping styles and social support

The more ConHD patients cope with problems by a seeking diversion (palliative reaction), seeking social support or using reassuring thoughts, the more interactions of social support they are likely to experience. High experience of negative reactions and discrepancies between desired and received social support (especially concerning emotional support and social companionship) in ConHD patients is positively related to passive reaction patterns. Furthermore, high coherence was found between appreciation by others and active problem solving as well as between daily emotional support and seeking social support.

One might expect that ConHD females, who cope with problems by passive reaction patterns more often than the reference group, are more likely to have negative experiences concerning social support. Because of lower active problem solving and lower seeking of social support compared to the reference group, ConHD females also seem likely to receive less appreciation shown by others and less emotional support respectively. On the contrary, coping styles in ConHD males seem to be favourably related to social support.

Unlike these hypotheses, both ConHD males and females reported less discrepancies in social support compared with the reference group. One of the reasons for this inconsistency in findings concerning the relationship between coping styles and social support, might be the underreporting of discrepancies in social support by overcompensation or denial mechanisms as suggested earlier. Another reason might be the use of different reference groups for coping styles and social support, since normative data were derived from the manuals of the questionnaires. Also, coping styles and social support might well be related to each other without resulting in less favourable social support in ConHD patients compared to the norm. Differences in coping styles between the ConHD females and reference females might not be large enough (all except for Active Solving of Problems not significant after Bonferroni correction), to represent differences in social support. Although ConHD females reported palliative reactions and reassuring thoughts in the normal range and less seeking social support compared to reference females, they still obtained higher scores on all of these coping styles, which were positively related to social support, compared to ConHD males. The overall more favourable coping styles regarding social support in females in general might therefore compensate small differences in coping styles between ConHD- and reference females.

In comparison to reference groups, ConHD females seem to have less favourable coping mechanisms than ConHD males. These results are not in line with the study of Salzer-Muhar et al. (2002) who found a negative self-concept in ConHD boys, assumed to be related to interference of reduced physical ability with peer relationships. It is likely that the negative consequences for social interactions might impede coping with the ConHD. Difference in age ranges of our study sample (20-46 years) and that of Salzer-Muhar et al. (2002) (12-16 years) could explain the different findings. Probably, different limitations which are involved with ConHD in different life stages, have different impacts on males and females. It is likely that, during adolescence, ConHD males are hampered by restricted physical abilities in their peer relationships to a larger extent than ConHD females (Salzer-Muhar et al., 2002). In early adulthood, ConHD females might be troubled by disease-related uncertainties concerning relationships and offspring more than ConHD males (van Rijen et al., submitted b). This age-linked difference between sexes is confirmed by Cyranowski et al. (2000), who states that in preadolescence, boys are more likely to become depressed than girls, while girls are more likely than boys to become depressed thereafter, assuming gender-linked vulnerability to life events as the underlying cause.

The relationship between coping styles and social support, should be considered as reciprocal rather than causal. Schmidt et al. (2002), in a study of attachment and coping with chronic disease in adult patients, states that secure attachment might be considered as an important inner resource in the emotional adaptation to chronic diseases. Insecure attachment was assumed to be related to less flexible coping. Rietveld et al. (2002) found that negative thoughts in adults with ConHD have a negative influence on psychosocial adjustment.

Limitations

The present patient sample contains a selection of five diagnostic categories and may therefore not be completely representative of all the congenital heart disease anomalies. The coping styles as measured in this study apply to problems in general and might not represent specific coping mechanisms that adult patients might use to deal with the congenital heart disease (e.g. denial of overcompensation).

Implications

Overall, in normative groups, females appear to seek and receive social support to a higher extent than males. This is also true for ConHD females. Remarkably, ConHD females seem to have less favourable coping styles than reference females, whereas ConHD males seem to have more favourable coping styles than reference males. In contrast, ConHD females did not report less favourable social support compared to reference females. Coping styles in ConHD patients should nevertheless be given special attention, since they are likely to determine the adaptation to the ConHD (Rietveld et al., 2002; Salzer-Muhar et al., 2002; Schmidt et al., 2002). Especially the significant less active problem solving in ConHD females is a point of concern, since passivity seems to imply less favourable social support. Intervention aimed at replacing passive coping styles by more active ones might increase a sense of control in ConHD patients. This might help especially ConHD females in dealing with problems involved with ConHD more adequately.

5 | Present subjective health status and longitudinal psychological well-being over a 10 year period in a cohort of adults with congenital heart disease

E.H.M. van Rijen
E.M.W.J. Utens
J.W. Roos-Hesselink
F.J. Meijboom
R.T. van Domburg
J.R.T.C. Roelandt
A.J.J.C. Bogers
F.C. Verhulst

Chapter 5

Present subjective health status and longitudinal psychological well-being over a 10 year period in a cohort of adults with congenital heart disease

Abstract

Aims: *Both medical status and subjective health experience determine the well-being of adult patients with congenital heart disease. We examined their present subjective health status and longitudinal course of psychological well-being.* ***Methods and results:*** *This study concerns the second psychological and medical follow-up of a cohort of 362 consecutive patients operated for congenital heart disease between 1968-1980 (age: 20-46 years). Present subjective health status and longitudinal course of psychological well-being from first to second follow-up were examined with the SF-36 and the Heart Patients Psychological Questionnaire respectively. The patient sample showed less favourable subjective health status concerning physical functioning, but more favourable social functioning, bodily pain and role limitations due to emotional problems, compared to a reference group. Younger female patients reported more role limitations due to physical functioning than older female patients. Patients with transposition of the great arteries showed a trend on subjective health status in the negative direction, but on the contrary reported the least bodily pain. Within the patient sample, displeasure increased, while social inhibition decreased over a 10-year period.* ***Conclusion:*** *Subjective health experiences of young female patients need extra attention. Patients with transposition of the great arteries overall seem to experience poorer physical health.*

Introduction

As a result of great improvements in surgical intervention and medical treatment of congenital heart disease over the last decades, the number of patients with congenital heart disease who survive into adulthood has increased enormously. Since this patient population of adults with congenital heart disease can be considered as rather new in medical practice, their health status self-evidently is of great interest (Meijboom et al., 1993; Meijboom et al., 1995; Meijboom et al., 1996; Meijboom et al., 1994; Roos-Hesselink et al., 2003). However, health status should not only be measured by medical standards. The way patients themselves experience and evaluate their health subjectively can be very determining for their well-being (Fekkes et al., 2001; Kamphuis et al., 2002; Ternestedt et al., 2001).

This phenomenon is generally referred to as “Quality of Life”. The World Health Organisation’s (WHO; World Health Organisation, 1948) definition of Quality of Life comprises the areas of physical, mental and social well-being. In the concept of Quality of Life, distinction should be made between subjective health status (patient’s perception of his or her state of abilities) versus health-related quality of life (patient’s personal feelings about these state of abilities) (Kamphuis et al., 2002). The present study focuses on subjective health status, since a concise and internationally well-known instrument for this aspect of Quality of Life was available, which enhanced the comparability of this study to other studies. At the start of this study, no validated Dutch instrument to measure health-related quality of life in adults with congenital heart disease was yet available.

In previous research, subjective health status in patient populations with severe as well as mild congenital heart disease has been examined. In a Quality of Life study among adults with complex congenital heart disease, Kamphuis et al. (2002) found a significantly worse subjective health status, compared to the general population, concerning physical functioning, role limitations due to physical functioning, vitality and general health perceptions. Greenwood (2000) found similar results in a group of adults who had undergone repair for a complex congenital cardiac lesion. In a study of Gersony et al. (1993), approximately 90% of adults with relatively mild congenital heart disease reported to be in good or excellent health. Fekkes et al. (2001) found that adults with mild congenital heart disease who experienced no social impediments reported a subjective health status comparable to the normal population. In contrast, mild congenital heart disease adults who did experience social impediments reported poorer subjective health status on physical functioning and general health perceptions compared to the general population.

The role of severity of cardiac diagnosis on subjective health status, however, seems ambiguous. Ternestedt et al. (2001) examined Quality of Life in a relative small sample of two diagnostic categories of congenital heart disease, 20 and 30 years postoperatively. Ternestedt found that patients with tetralogy of Fallot rated their Quality of Life higher than did patients with generally considered less severe atrial septal defect. Also, more patients with atrial septal defect than patients with tetralogy of Fallot stated that the congenital heart disease had a negative impact on their adult life. In an adult patient sample comprising several congenital cardiac defects, Lane et al. (2002) found poorest subjective health status among patients with inoperable and cyanotic conditions and paradoxically, in those deemed surgically cured. Unfortunately, the response rate in this study was low.

Until now, limited research has been executed in consecutive series of patients with congenital heart disease. In the present study, a cohort of patients with congenital heart disease was examined, in which subjective health status could be compared between diagnostic categories directly. Moreover, little is known about the longitudinal course of psychological well-being in adults with congenital heart disease. In the present study, besides the subjective health status at present, the related concept of psychological well-being was examined longitudinally in a large selection of the cross-sectional cohort, in order to provide more insight in how the way adult patients are affected with the congenital heart disease changes over time.

Subjective health status was examined in a large consecutive series of patients, who underwent open heart surgery for congenital heart disease between 1968 and 1980, encompassing 5 diagnostic categories with a large age range in adulthood and a high response rate. Only a miscellaneous diagnostic group was excluded. The aims of the present study were:

1. to compare *the present subjective health status* of adults with operated congenital heart disease with that of the normal population.
2. to identify the role of sex, age and cardiac diagnosis on the present subjective health status in adults operated for congenital heart disease.
3. to examine *the longitudinal course of psychological well-being* over a period of 10 years in adults with operated congenital heart disease.
4. to identify the role of sex and cardiac diagnosis on the longitudinal course of psychological well-being over a period of 10 years in adults with operated congenital heart disease.

Methods

Inclusion criteria

During the first follow-up of this study, which took place in 1989-1991, all consecutive patients who underwent their first open heart surgery for congenital heart disease between 1968 and 1980 in the Erasmus MC, and who were younger than 15 years at the time of surgery, were eligible. This population is described in detail elsewhere (Utens et al., 1998a; Utens et al., 1994; Utens et al., 1993).

The target population of the second follow-up (2000-2001) consisted of the 498 patients of the first follow-up. From the 498 patients we excluded 61 patients who belonged to a miscellaneous diagnostic group, consisting of small numbers of patients with a variety of congenital heart defects (aortic stenosis, discrete subaortic stenosis, atrioventricular septal defect, pulmonary atresia, tricuspid atresia, total abnormal pulmonary venous drainage, truncus arteriosus, and further miscellaneous diagnoses). Furthermore we excluded 11 deceased patients, 26 untraceable patients and 1 patient who had undergone a heart transplantation.

Patient sample

Of the 399 eligible patients, 37 refused to participate. The remaining 362 patients participated in the psychological examination of this study. The overall response rate, corrected for deceased persons and persons lost to follow-up was 90.7%.

Subjective health status. Of the 362 participating patients, 11 mentally retarded patients and 1 patient with a language problem were not able to complete the questionnaire concerning subjective health status, 1 patient who participated by mail and phone did not return it. This resulted in usable questionnaires for 87.5% of the eligible patient sample. The present sample of respondents on subjective health status consisted of 349 patients (189 males, 160 females, mean age 30 years, age range 20-46 years). Numbers of patients belonging to different diagnostic groups were: closure of atrial septal defect (n=93), closure of ventricular septal defect (n=92), corrective surgery for tetralogy of Fallot (n=72), Mustard procedure for transposition of the great arteries (n=55) and surgery for pulmonary stenosis (n=37). The mean ages in these diagnostic groups were 33, 30, 30, 26 and 32 years respectively. Medical history and physical condition of these patients at the time of the first and second follow-up are described elsewhere (Meijboom et al., 1993; Meijboom et al., 1995; Meijboom et al., 1996; Meijboom et al., 1994; Roos-Hesselink et al., 2003).

Psychological well-being. At the second follow-up, 8 mentally retarded patients and 1 patient with a language problem were not able to complete the questionnaire regarding psychological well-being, 1 patient who participated by mail and phone did not return it, another patient did not complete it entirely. This resulted in usable questionnaires for 88.0% of the eligible patient sample (n=351). Because of longitudinal comparisons, only patients for whom data regarding psychological well-being were available at both the first and second follow-up, were included. These data were available for 243 patients (125 males, 118 females; mean age 33 years, age range 26-46 years; atrial septal defect: n=86, ventricular septal defect: n=54, tetralogy of Fallot: n=48, transposition of the great arteries: n=24, pulmonary stenosis: n=31). None of the patients younger than 26 years at present were included, since these patients did not complete the questionnaire concerning psychological well-being at the first assessment, because it was not fitted for their age ranges then.

Reference group

Subjective health status. Normative data were derived from a nation wide, population-based Dutch health status survey (Aaronson et al., 1998). Its total sample contained 1742 participants with an age range of 16-94 years (56% male and 44% female). A selection of this sample, i.e. participants with an age range of 16-40 years, was used as reference group for the present study.

Psychological well-being. Since longitudinal comparisons were performed within the congenital heart disease patient sample, no reference data were used. The original purpose of the questionnaire regarding psychological well-being was to assess well-being in adult cardiac patients, the vast majority suffering from acquired heart disease. No recent age-compatible normative data from the general population were available.

Instruments

Subjective health status was assessed by the SF-36 Health Survey (SF-36; Ware & Sherbourne, 1992). The SF-36 contains 36 questions with standardised response choices. Subjective health status is measured on 8 scales: Physical functioning (performing all sorts of physical activities), Role limitations due to physical health problems (problems with work or other daily activities as a result of physical health), Social functioning (interference with normal social activities due to physical and emotional problems), Bodily pain, General mental health, Role limitations due to emotional problems (problems with work or other daily activities as a result of emotional problems), Vitality (feeling either tired/ worn out all the time versus full of energy) and General health perceptions (believing personal health is either poor and likely to get worse versus excellent). A standard version of the SF-36 was administered, which asks patients to fill in the questionnaire as applicable to the 4 weeks prior to the psychological examination. Good reliability and validity for the Dutch version of the SF-36 has been reported (Aaronson et al., 1998).

Psychological well-being was assessed by the Heart Patients Psychological Questionnaire (Erdman, 1982), which contains 52 items. The response categories are: “true”; “?’; “not true”. Psychological well-being is measured on 4 scales: Well-being (feeling happy, satisfied and relaxed), Feelings of being disabled (experienced discrepancy between desired and real abilities), Displeasure (negative mood and emotions) and Social inhibition (feeling unpleasant among other people). Satisfactory reliability and validity for the Heart Patients Psychological Questionnaire have been reported (Erdman, 1982).

Assessment procedures

All patients were traced, approached uniformly and signed an informed consent before participating. During their visit to the Erasmus MC, patients were interviewed and tested by a psychologist (EvR) and medically examined by a cardiologist. Some included patients (n=28) could not visit the hospital for practical or emotional reasons; they filled in the questionnaires at home and returned it by mail.

Statistical analyses

One sample t-tests, based on 95% confidence intervals, were used to test differences between the patient sample and reference group on the SF-36. To identify the role of sex (male, female), age (20-29 years, 30-46 years) and cardiac diagnosis (atrial septal defect, ventricular septal defect, tetralogy of Fallot, transposition of the great arteries and pulmonary stenosis) within the congenital heart disease patient sample, univariate analyses of variance (ANOVAs), based on 95% confidence intervals, were performed on all SF-36-scales. The longitudinal comparisons on the Heart Patients Psychological Questionnaire were analysed in a One Factor Repeated Measures Design, with sex and diagnosis as within-subject variables. The variable age was left out here, because no patients younger than 26 years were included. In order to correct for multiple comparisons in analyses mentioned above, Bonferroni corrections were applied. Since, after Bonferroni correction, socioeconomic status (low, middle high) showed significant main effects on only 2 out of the total 12 scales of both questionnaires and no significant differences in socioeconomic status between diagnostic categories were found, socioeconomic status was not corrected for.

Results

Subjective health status

Table 5.1 shows the mean scores on the SF-36 of the congenital heart disease patient sample and an age compatible reference group from the normal population. The patient sample showed less favourable Physical functioning compared to the reference group, indicating more impairment for patients with congenital heart disease on physical activities. The patient sample obtained more favourable scores on Social functioning, Bodily pain and Role limitations due to emotional problems compared to the reference group. This means that patients with congenital heart disease reported less experience of bodily pain, less interference with normal social activities due to physical and emotional problems and less problems with work or other daily activities as a result of emotional problems, compared to a reference group from the normal population. These effects remained significant after Bonferroni correction for 8 comparisons.

Table 5.1 Mean scores of the ConHD patient sample and reference group on the SF-36 scales

	ConHD patient sample (N = 349)	Reference group	p-value
Physical functioning	90.3	93.1	$< 0.001^{a,b}$
Role limitations due to physical functioning	89.0	86.4	0.064
Social functioning	90.9	87.8	$< 0.001^{a,b}$
Bodily pain	85.6	80.9	$< 0.001^{a,b}$
General mental health	80.0	78.7	0.096
Role limitations due to emotional problems	90.1	85.4	$0.001^{a,b}$
Vitality	70.7	70.7	0.998
General health perceptions	76.8	78.2	0.184

Note. Abbreviation used: ConHD = Congenital Heart Disease.
The SF-36 scales range from 0 to 100. Lower scores indicate poorer subjective health status, higher scores indicate more favourable subjective health status.
[a] Significant at 95% confidence level, $p < 0.05$.
[b] Significant after Bonferroni correction for 8 comparisons, $p < 0.006$.

Effects of sex, age and diagnosis

ANOVA’s on SF-36 scale scores within the congenital heart disease patient sample revealed significant main effects for sex on Physical functioning, Social functioning, General mental health and Vitality, all indicating more favourable scores for males compared to females. All effects, except for the effect on Physical functioning, did not remain significant after Bonferroni corrections for 8 comparisons.

No significant main effect for age was found. A significant interaction effect for sex x age was found on Role limitations due to physical functioning ($p = 0.033$). Younger congenital heart disease males (20-29 years) scored more favourably on Role limitations due to physical functioning than older congenital heart disease males (30-46 years), whereas younger congenital heart disease females reported less favourable Role limitations due to physical functioning compared to older congenital heart disease females. This interaction effect did not remain significant after Bonferroni correction for 8 comparisons.

Table 5.2 shows the mean scores on the SF-36 scales for the different cardiac diagnostic groups. No significant main or interaction effect for cardiac diagnosis was found.

Table 5.2 Mean scores of different cardiac diagnostic groups within the ConHD patient sample on the SF-36 scales

	ASD (N = 93)	VSD (N = 92)	ToF (N = 72)	TGA (N = 55)	PS (N = 37)	p-value
Physical functioning	92.1	91.7	88.5	86.0	91.9	0.11
Role limitations due to physical functioning	89.8	86.4	88.2	87.3	97.3	0.25
Social functioning	90.3	91.0	92.5	89.1	91.9	0.86
Bodily pain	83.9	84.6	88.0	89.7	81.9	0.57
General mental health	80.3	79.6	81.3	79.2	79.5	0.93
Role limitations due to emotional problems	90.7	89.9	90.3	86.7	93.7	0.67
Vitality	71.4	69.9	73.3	67.8	70.0	0.19
General health perceptions	79.9	77.1	75.8	72.9	76.3	0.14

Note. Abbreviations used: ConHD = Congenital Heart Disease, ASD = atrial septal defect, VSD = ventricular septal defect, ToF = tetralogy of Fallot, TGA = transposition of the great arteries, PS = pulmonary stenosis. The SF-36 scales range from 0 to 100. Lower scores indicate poorer subjective health status, higher scores indicate more favourable subjective health status.

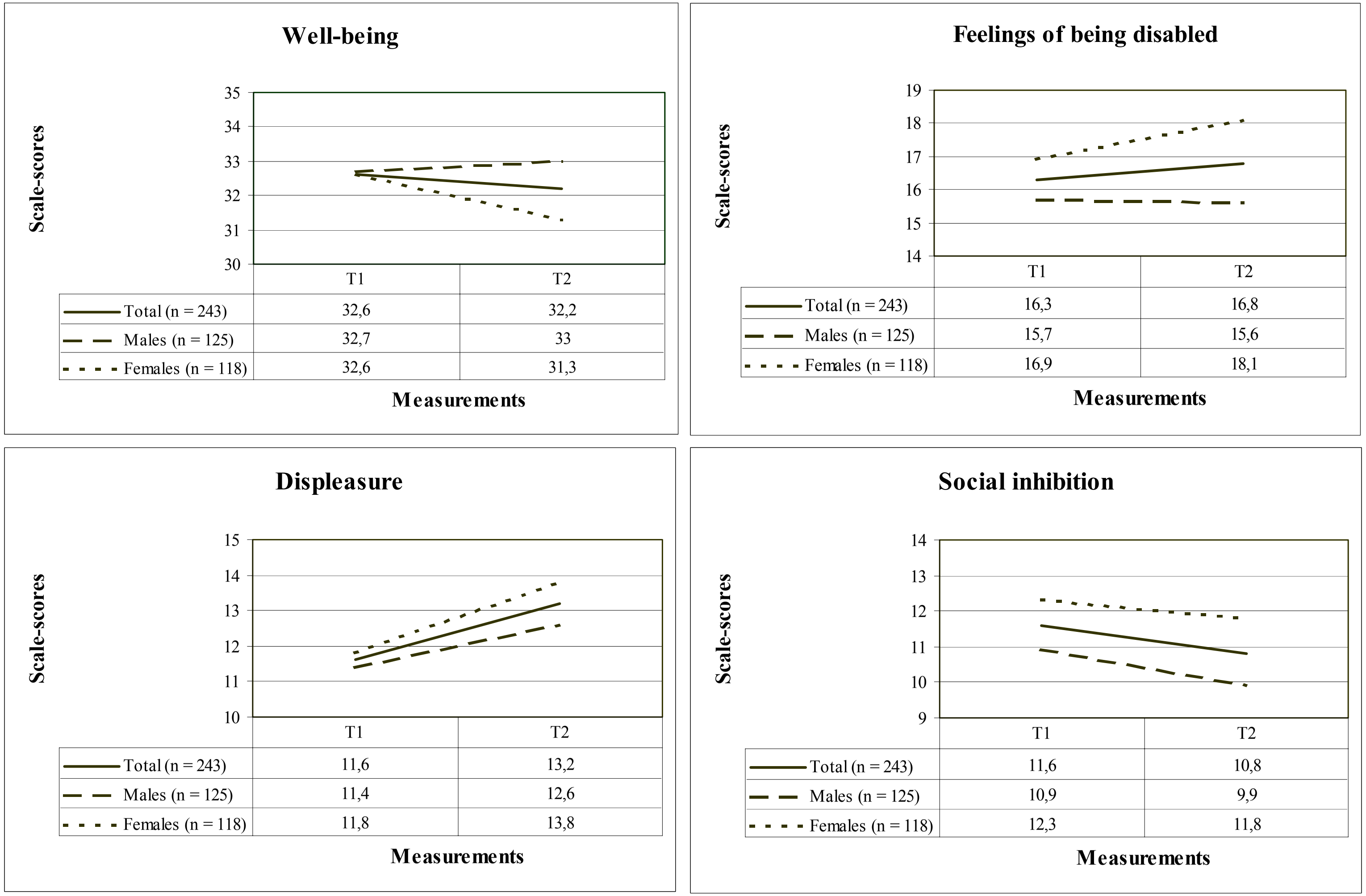
Longitudinal course of psychological well-being

Repeated Measures analyses on Heart Patients Psychological Questionnaire scale scores within the congenital patient sample revealed significant change over time between the first and second follow-up on Displeasure and Social Inhibition. Over this 10-year period, Displeasure increased and Social inhibition decreased within the congenital patient sample. Both of these effects, remained significant after Bonferroni correction for 4 comparisons. Figure 1 shows the changes over time on the Heart Patients Psychological Questionnaire scales for the total congenital heart disease patient sample, as well as for males and females separately.

Effects of sex and diagnosis

A significant effect over time for sex was found on Well-being. Congenital heart disease males showed an increase in Well-being over a 10-year period of time. In contrast, congenital heart disease females showed a decrease in Well-being. This effect did not remain significant after Bonferroni correction for 4 comparisons. No significant effects for diagnosis were found.

Figure 5.1 Longitudinal course of ConHD patient sample (age range 26-46 years) on HPPQ-scales



Note. Abbreviations used: ConHD = Congenital Heart disease, HPPQ = Heart Patient Psychological Questionnaire.
Values on axis Measurements: T1 = Follow-up time 1 (1989-1991), T2 = Follow-up time 2 (2000-2002).
On Well-being, high scores indicate favourable psychological well-being. On Feelings of being disabled, Displeasure and Social inhibition, high scores indicate poor psychological well-being.

Discussion

Subjective health status

Despite higher impairment on physical functioning, patients with congenital heart disease reported to be less bothered by physical and emotional problems in social activities, to have less problems with work or other daily activities as a result of emotional problems and to experience less bodily pain, compared to a reference group from the normal population.

The more favourable results on Social functioning and Role limitations due to emotional problems, which imply less interference of emotional and/ or physical problems on daily life, might indicate adequate coping with the disadvantageous consequences of the congenital heart disease. However, overcompensation and denial of problems associated with the congenital heart disease have also been suggested as coping strategies in adult patients with congenital heart disease (Ternestedt et al., 2001; Utens et al., 1996; Utens et al., 1994). Indications for denial mechanisms in adults with congenital heart disease were previously described regarding sick-leave and psychopathology for the present cohort (van Rijen et al., 2003; van Rijen et al., submitted b). Except for less active problem solving in congenital heart disease females, coping styles in adults with congenital heart disease in general seemed similar to those of the normal population (van Rijen et al., submitted a). It thus seems difficult to distinguish between adequate coping (not being bothered by physical and/ or emotional problems too much) versus denial of problems and their consequences. Nevertheless, since poorer physical functioning was reported here, overall wishful thinking and exaggerating positive outcome do not seem the most likely explanations. Possibly, when growing up with congenital heart disease, these adult patients have learned how not to restrain themselves too much by emotional or physical problems. This is in line with results previously found in the present cohort, namely the overall capacity of leading normal lives and motivation to make good use of their abilities (van Rijen et al., 2003).

Since higher impairment on physical functioning was reported in the present study and higher reporting of somatic complaints was previously described for this same cohort (van Rijen et al., submitted b), the low experience of bodily pain can be considered remarkable. In a study of adults with univentricular hearts, Saliba et al. (2001) found favourable outcome on pain items, while at the same time physical problems were reported. In a study of adults from a large spectrum of various cardiac diagnoses, Lane et al. (2002), using the same instrument for subjective health status (SF-36), only found less favourable experience of bodily pain compared to the normal population in those patients with the worse (inoperable) conditions. Experiences of treatment and surgery might have lead to higher pain thresholds in patients with congenital heart disease. In judging their experience of pain, adults with congenital heart disease might use different frames of reference than the normal population. This assumption is strengthened by the finding that, though no significant differences over all diagnostic categories were found, the generally considered most severely affected patients with tetralogy of Fallot and transposition of the great arteries obtained by far the most favourable scores for experience of bodily pain. Presumably, medical experiences put bodily pain in a different perspective.

Effects of sex, age and diagnosis

Congenital heart disease males reported better performing on physical activities, less interference with normal social activities due to physical and emotional problems, better mental health and better vitality compared to congenital heart disease females. These results are in line with differences between sexes found in the normal population (Aaronson et al., 1998).

Psychological examination of the present cohort previously revealed higher levels of psychopathology among young congenital heart disease females compared to a reference group (van Rijen et al., submitted b). As an explanation for this finding, it was suggested disease-related uncertainties concerning sexual relationships and offspring might have a great impact on the lives of young congenital heart disease females, since these issues involve their own bodily functioning. This might also explain why, in the present study, younger congenital heart disease females reported more limitations due to physical health problems than older congenital heart disease females. Older congenital heart disease females might feel less restricted by physical problems once they have overcome these uncertainties and a stable relationship and family life are established.

Although no significant effect for diagnosis was found, it can be noticed that, when compared to the other diagnostic categories, the patients with transposition of the great arteries reported the poorest subjective health status on nearly all scales, except for limitations due to physical functioning and experienced bodily pain. In accordance with previous studies concerning subjective health status in adults with complex congenital heart disease (Greenwood, 2000; Kamphuis et al., 2002), the poorest outcome for patients with transposition of the great vessels compared to other diagnostic categories was found on Physical functioning, Vitality and General health perceptions. This might indicate that patients with transposition of the great vessels do experience some disadvantageous consequences of the congenital heart disease on their physical health.

Longitudinal course of psychological well-being

Over a 10-year period of time, decline as well as improvement were observed in the course of psychological well-being in congenital heart disease patients. Feelings of displeasure (negative mood and emotions) increased, while social inhibition (feeling unpleasant among other people) decreased over time. This means that, in the transition from young to middle adulthood, though negative emotions might become somewhat more apparent, adults with congenital heart disease feel more secure in social situations as they grow older. Since the increase in displeasure was located near the lower limit of the scorings range, it might also be partly attributed to regression to the mean.

Effects of sex and diagnosis

Figure 1 shows that at both follow-up 1 and 2, congenital heart disease females scored less favourably on all of the Heart Patients Psychological Questionnaire scales compared to congenital heart disease males. Only concerning well-being, the change over time can be considered different for both sexes, since congenital heart disease males reported increase in well-being, whereas congenital heart disease females reported decrease in well-being.

Remarkably, for the different diagnostic categories, no significant differences in longitudinal course of psychological well-being were found, not even for feelings of being disabled.

Conclusions

Despite poorer physical functioning, overall positive outcomes were found for subjective health status in the congenital heart disease patient sample, since further results were similar or more positive compared to the reference group. Although negative emotions increased somewhat during adulthood, congenital heart disease patients became more secure in social situations.

Congenital heart disease females showed less favourable subjective health status and course of psychological well-being compared to congenital heart disease males. Although most differences between sexes were comparable to those in the normal population, it is striking that experience of limitations due to physical health problems diminish as congenital heart disease females grow older. This strengthens the assumption that disease-specific uncertainties might trouble young congenital heart disease females. In clinical practice, attention should thus be paid to the way young congenital heart disease females experience and evaluate their health subjectively. If needed, counselling should be provided.

Sample sizes of the different diagnostic categories might not have been large enough to reveal differences between them. Compared to other diagnoses, patients with transposition of the great arteries showed a trend on subjective health status in the negative direction. In contrast, they seem to report the least bodily pain. Regarding these contradicting subjective evaluations of health status, we conclude that in clinical practice this group of patients deserves special attention. Besides higher pain thresholds or different evaluation of pain, this finding might also indicate overcompensation.

6 | Medical predictors for psychopathology in adults with operated congenital heart disease

E.H.M. van Rijen
E.M.W.J. Utens
J.W. Roos-Hesselink
F.J. Meijboom
R.T. van Domburg
J.R.T.C. Roelandt
A.J.J.C. Bogers
F.C. Verhulst

Chapter 6

Medical predictors for psychopathology in adults with operated congenital heart disease

Abstract

Aims: *To test the predictive value of medical variables, covering the complete medical course from birth up till now, for long-term behavioural and emotional problems in adulthood, in patients operated for congenital heart disease in childhood.* ***Methods and results:*** *This study concerns the second psychological and medical follow-up of a cohort of patients operated for congenital heart disease (n=362; age 20-46 years). Behavioural and emotional problems were assessed with the Young Adult Self-Report and the Young Adult Behavior Checklist. Medical predictor variables were derived from medical examination and file search. Being female, low exercise capacity and restrictions imposed by physicians are significant predictors for behavioural and emotional problems as reported by patients themselves. Regarding the scar, subjective experiences of patients form a better predictor for later problems than objective judgements by physicians. Early hospitalisations with reoperations are predictive for behavioural and emotional problems as reported by other informants. The cardiac diagnoses ventricular septal defect and transposition of the great arteries are associated with higher levels of behavioural and emotional problems.* ***Conclusion:*** *Recent experiences concerning the scar, physical condition and imposed restrictions are the strongest predictors for behavioural and emotional problems as reported by patients themselves.*

Introduction

As a result of increasing survival rates for children with congenital heart disease over the last decades, a new patient population of adults with congenital heart disease is emerging today. Little is known about behavioural and emotional problems of congenital heart disease adults, who bear the consequences of not only their medical condition, but also of growing up with a congenital anomaly. Studies on the level of psychopathology in congenital heart disease adults show conflicting results with some showing elevated levels of psychopathology (Brandhagen et al., 1991; van Rijen et al., submitted b) and others showing levels similar to those of peers (Cox et al., 2002; Utens et al., 1998a). Some studies identified predictors for behavioural and emotional problems in children with congenital heart disease, such as maternal perceptions (DeMaso et al., 1991), circulatory arrest (Bellinger et al., 1997; Utens et al., 1998b) and number of heart operations (Utens et al., 1998b). Medical predictors of long-term psychopathology in congenital heart disease adults are yet to be explored.

This study is part of the second follow-up conducted in 2000 and 2001 of a multidisciplinary study which provides longitudinal psychological and medical data, of patients with congenital heart disease, 20 to 33 years after their first cardiac operation. The first follow-up conducted in 1989 through 1991 contained a prediction study on behavioural and emotional problems in children and adolescents with congenital heart disease (Utens et al., 1998b). Since all patients had reached adulthood at the time of the second follow-up, we were able to perform a similar prediction study on a large sample of, now all adult, congenital heart disease patients.

Investigating predictors for psychopathology in congenital heart disease adults could be helpful for identifying individuals at risk. Also, aspects in the treatment and support of congenital heart disease adults which need special attention could be specified. The main aim of this study was to determine to what extent variables concerning: [1] biographical status, [2] medical history, [3] first open heart surgery and direct post-operative course, [4] medical course before 1990, [5] medical course after 1990, [6] present contacts with physicians, and [7] present medical status, can predict long-term behavioural and emotional problems in adulthood.

Method

Inclusion criteria

During the first follow-up of this study, which took place in 1989-1991, all consecutive patients who underwent their first open heart surgery for congenital heart disease between 1968 and 1980 in the Erasmus MC, and who were younger than 15 years at the time of surgery, were eligible. For a detailed description of this population, see Utens et al. (Utens et al., 1998a; Utens et al., 1994; Utens et al., 1993).

The target population of the present follow-up (2000-2001) consisted of the 498 patients of the first follow-up. From the 498 patients, 61 patients who belonged to a miscellaneous diagnostic group, consisting of small numbers of patients with a variety of congenital heart defects (aortic stenosis, discrete subaortic stenosis, atrioventricular septal defect, pulmonary atresia, tricuspid atresia, total abnormal pulmonary venous drainage, truncus arteriosus, and further miscellaneous diagnoses) were excluded. Furthermore, 11 patients had died, 26 patients were untraceable and 1 patient had undergone a heart transplantation.

Patient sample

Of the remaining 399 patients, 37 refused to participate. Among the participating 362 patients, 10 mentally retarded patients were unable to complete the self-report questionnaire and 3 questionnaires were not usable because of incomplete information, resulting in 349 self-reports. Regarding other informant reports, of the 362 participating patients, for 1 mentally retarded patient the questionnaire was not applicable, 3 patients did not have a suitable other informant because of language problems, 19 questionnaires were not filled in and 5 questionnaires were not usable because of incomplete information, resulting in 334 other informant reports. The overall response rate, corrected for deceased persons and persons lost to follow-up was 90.7%, resulting in usable self-reports and other informant reports for respectively 87.5% and 83.7% of the eligible patient sample.

The part of the patient sample for which self-reports were available consisted of 188 males and 161 females (mean age: 30 years, age range 20-46 years). Numbers of patients belonging to each diagnostic group were: closure of atrial septal defect (ASD; n=93), closure of ventricular septal defect (VSD; n=92), corrective surgery for tetralogy of Fallot (ToF; n=72), Mustard procedure for transposition of the great arteries (TGA; n=55) and surgery for pulmonary stenosis (PS; n=37). The mean ages in these diagnostic groups were 33, 30, 30, 26 and 32 years respectively. Medical history and physical condition of these patients at the time of the first follow-up (Meijboom et al., 1993; Meijboom et al., 1995; Meijboom et al., 1996; Meijboom et al., 1994) and second follow-up (Roos-Hesselink et al., 2003) are described elsewhere.

Outcome variables

Psychopathology in young adults with congenital heart disease was assessed with a self-report questionnaire: the Young Adult Self-Report (YASR; Achenbach, 1997) and a parallel questionnaire to be completed by significant others, such as a parent or a spouse: the Young Adult Behavior Checklist (YABCL; Achenbach, 1997).

Good reliability and validity of the YASR (Achenbach, 1997) have been confirmed for an earlier Dutch version of the YASR (Ferdinand et al., 1995). The YASR contains 110 problem items. The response format is 0 = “not true”, 1 = “somewhat or sometimes true”, and 2 = “very true or often true”. The YASR can be scored on 8 syndrome scales:

Anxious/Depressed, Withdrawn, Somatic Complaints, Thought Problems, Attention Problems, Intrusive Behavior, Delinquent Behavior, and Aggressive Behavior, and two broad-band groupings of syndromes: Internalizing (consisting of the Anxious/Depressed and Withdrawn scales) and Externalizing (consisting of Intrusive Behavior, Delinquent Behavior and Aggressive Behavior scales). A Total Problems score is computed by summing the individual item scores. High scores indicate the presence of behavioural and emotional problems and thus are unfavourable. Low scores are favourable.

The YABCL (Achenbach, 1997), a parallel version of the YASR, is to be completed by an informant who is familiar with the young adult, including a partner, parent or other close relative or friend. The YABCL contains 105 problem items and consists of the same scales as the YASR. Good reliability and validity was reported for the YABCL (Achenbach, 1997).

The YASR and YABCL were originally developed for 18- to 30 year-olds. However, since normative data were not used in this prediction study, we decided to include the complete patient sample. For this particular study, only the scales Internalizing, Externalizing and Total problems of both YASR and YABCL were used.

Prediction variables

In order to examine the predictive value of medical variables on psychopathology in congenital heart disease adults, 7 clusters of prediction variables were chosen on theoretical and/or medical grounds. Data were partly derived from a retrospective medical file search and medical examination by a cardiologist during the first follow-up (F.M.), and partly derived form medical examination by a cardiologist during the second follow-up (J.R.).

The variable ‘date first open heart surgery’ was converted to the number of days that elapsed since the first patient of this cohort received open heart surgery. This was done to examine accurately the impact of the exact period in which the first surgery took place, since surgical techniques developed at high pace. The variable ‘postoperative course’ was dichotomised: 0 = no complications, 1 = with complications. The variable ‘results operation’ was dichotomised: 0 = good, 1 = moderate or poor. Most dichotomous prediction variables were coded as: 0 = favourable or risk absent and 1 = unfavourable or risk present, except for the prediction variables ‘sex’ (0 = female, 1 = male) and ‘ECG sinus rhythm’ (0 = sinus rhythm absent, 1 = sinus rhythm present). The maximum exercise capacity (derived from bicycle ergometry) is the percentage from what might be expected considering age, sex and body weight, compared to the normal Dutch population.

Assessment procedures

All patients were traced, approached uniformly and signed an informed consent before participating. During their visit to the Erasmus Medical Center Rotterdam, patients were interviewed and tested by a psychologist (EvR) and medically examined by a cardiologist. The YASR was filled in by the patient during the psychological examination. Some patients (n = 28) who could not visit the hospital for practical or emotional reasons completed the YASR at home and returned it by mail. The YABCL was filled in at home, by someone familiar to the patient (partners, spouses: 64%, parents: 32%, siblings, close friends and others: 4%) and was returned by mail. For patients, who were accompanied during their hospital visit by a familiar person (e.g. parent, spouse), the YABCL was completed in the waiting room.

Statistical Analyses

A three-phase strategy was followed, for each of the six outcome measures. Linear regression analysis was applied with only main terms entered in the regression model. In phase 1, each of the separate prediction variables was tested on the YASR and YABCL outcomes (univariate analysis). This was done to explore the predictive quality of each predictor separately. In phase 2, each cluster (i.e. combination) of predictors was related to the YASR and YABCL outcomes (multivariate analysis). The following clusters were used: biographical status, medical history, first open heart surgery and direct post-operative course, medical course before 1990, medical course after 1990, present contacts with physicians and present medical status. Since this phase served as a first, broad selection of predictors, the p-values were set to levels of 0.20 (backward elimination procedure). Variables which remained in the regression model, applied to the clusters of prediction variables, were candidate predictors for the final model. In order to correct for sex- and age effects, sex and age were forced into each analysis in phase 1 and 2. The final model in phase 3 contained all significant variables from phase 2 (p-values were set at 0.05, backward elimination procedure). Variables which showed significant results in this final model were regarded as the final predictors of YASR and YABCL outcomes. In the tables, the betas presented are standard regression coefficients which express the strength of the relationship between each prediction variable and the outcome variable. Plus versus minus indicates respectively the positive versus negative direction of the relation between the prediction variable and the outcome variable.

The distributions of the 6 outcomes were positively skewed, as could be expected since the YASR and YABCL can be regarded as instruments for assessing psychopathology.

In order to normalise the distributions, transformations were applied to each outcome by taking square roots on the problem scores added by 1 (since square roots can only be calculated on scores >0).

Results

Phase 1: univariate analyses

Table 6.1 shows the results of phase 1 analyses. 'Restrictions by scar experienced by patients (sometimes)' was the only variable which significantly predicted all six outcome measures. Being female significantly predicted higher (that is unfavourable) scores on both YASR and YABCL Internalizing and Total problems. The diagnostic category transposition of the great arteries was a significant predictor for all YASR scales. The diagnostic category ventricular septal defect was a significant predictor for YABCL Total problems. Reoperation(s) before 1990 was related to YABCL Internalizing and Total problems. Medical check-ups for the heart once a year or more were associated with higher scores on YASR Internalizing. Restrictions imposed by physician was related to higher scores on YASR Total problems. Poor physical condition (low maximum exercise capacity) was associated with higher scores on YASR Internalizing. Age at follow-up, hospitalisations without reoperation(s) before 1990 and having a pace-maker were associated with lower (that is favourable) scores on some outcome variables.

Phase 2: multivariate analyses

Since phase 2 analyses only served as a first selection of predictors and as an in-between model , the results are not presented here.

Phase 3: the final model

The results of the final prediction model are presented in Table 6.2. 'Restrictions by scar experienced by patients (sometimes)' was related to higher problem scores on nearly all outcome variables, except for YABCL Internalizing. Reoperation(s) before 1990 significantly predicted higher scores on all YABCL scales. Being female significantly predicted higher scores on YASR Internalizing and Total problems. Poor physical condition (low maximum exercise capacity) was associated with higher scores on YASR Internalizing and Total problems. The diagnostic categories ventricular septal defect and transposition of the great

Table 6.1 Prediction of outcomes on YASR and YABCL Internalizing, Externalizing and Total problems by seperate prediction variables

Prediction variables	YASR Internalizing	YASR Externalizing	YASR Total problems	YABCL Internalizing	YABCL Externalizing	YABCL Total Problems
Biographical status						
Sex (Male)	-0.227^{C}	0.027	-0.136^{a}	-0.166^{b}	-0.067	-0.136^{a}
Age, in years	0.016	-0.119^{a}	-0.048	0.029	-0.029	-0.028
Medical History						
Duration of pregnancy, in weeks	-0.022	0.098	0.053	-0.019	-0.005	-0.010
Weight at birth, in grams	-0.041	0.059	0.012	-0.006	-0.007	-0.019
Palliative surgery prior to surgical repair	0.068	0.039	0.079	0.069	0.015	0.054
Cardiac diagnosis (reference category: ASD)						
VSD	0.071	0.121	0.120	0.090	0.126	0.172^{a}
ToF	0.027	-0.002	0.044	0.031	0.035	0.093
TGA	0.154^{a}	0.147^{a}	0.178^{b}	0.128	0.039	0.093
PS	0.037	0.010	0.041	0.120	0.016	0.076
First open heart surgery and direct post-operative course						
Date first open heart surgery, in days	0.030	-0.098	-0.009	-0.106	-0.023	-0.090
Age first open heart surgery, in years	0.037	-0.082	0.012	-0.149	-0.021	-0.118
Postoperative course (with complications)	-0.007	-0.009	-0.001	0.010	0.002	0.030
Results operation (moderate or poor)	-0.007	0.016	0.011	0.003	0.005	0.023
Medical course before 1990						
Number of hospitalisations as a result of heart problems	0.028	-0.081	0.022	0.025	0.004	0.048
Course of hospitalisations as a result of heart problems (reference category: no hospitalisations)						
Hospitalisations without reoperation(s)	-0.009	-0.055	-0.007	-0.119^{a}	-0.128^{a}	-0.117^{a}
Hospitalisations with reoperation(s)	0.059	-0.005	0.064	0.125^{a}	0.069	0.114^{a}
Medical course after 1990						
Number of hospitalisations as a result of heart problems	0.047	-0.020	0.052	-0.043	0.009	0.003
Course of hospitalisations as a result of heart problems (reference category: no hospitalisations)						
Hospitalisations without reoperation(s)	0.085	-0.074	0.049	-0.064	-0.101	-0.111
Hospitalisations with reoperation(s)	-0.025	0.073	0.006	0.054	0.146	0.166
Present contacts with physicians						
Medical check-ups for the heart (reference category: no medical check-ups)						
Less than once a year	-0.044	-0.088	-0.065	-0.057	-0.024	-0.030
Once a year or more	0.116^{a}	-0.005	0.104	0.060	-0.030	0.042
Restrictions imposed by physician	0.090	0.087	0.125^{a}	0.001	0.093	0.069
Present medical status						
Medication for the heart	0.096	-0.038	0.053	0.063	-0.005	0.033
Scar judged by physician (reference category: well healed)						
Moderately healed	-0.035	-0.102	-0.079	-0.045	-0.091	-0.089
Poorly healed	-0.035	0.040	0.004	-0.048	0.022	-0.023
Restrictions by scar experienced by patients (reference category: never)						
Sometimes	0.254^{c}	0.135^{a}	0.216^{c}	0.112^{a}	0.140^{a}	0.137^{a}
Often	0.081	0.067	0.090	0.098	0.048	0.078
Pacemaker	-0.006	-0.074	-0.014	-0.055	-0.112^{a}	-0.085
Maximum exercise capacity in %	-0.110^{a}	-0.079	-0.108	-0.018	0.002	-0.019
ECG sinus rhythm	-0.033	0.030	-0.020	0.059	0.076	0.086

Note. Numbers in table are standardised coefficients β. Abbreviations used are YASR = Young Adult Self-Report, YABCL = Young Adult Behavior Checklist, ASD = Atrial Septal Defect, VSD = Ventricular Septal Defect, ToF = Tetralogy of Fallot, TGA = Transposition of the Great Arteries, PS = Pulmonary Stenosis.

[a] $p < 0.05$.

[b] $p < 0.01$.

[c] $p < 0.001$.

Table 6.2 Final results of prediction of YASR and YABCL Internalizing, Externalizing and Total problems by separate prediction variables

	ß	P	R^2
YASR Internalizing			
Sex (Male)	-0.201	< 0.001	0.118
Restrictions by scar experienced by patients (reference category: never)			
Sometimes	0.238	< 0.001	
Maximum exercise capacity in %	-0.105	0.044	
YASR Externalizing			
Cardiac diagnosis (reference category: ASD)			
VSD	0.119	0.036	0.065
TGA	0.148	0.009	
Scar judged by physician (reference category: well healed)			
Moderately healed	-0.137	0.013	
Restrictions by scar experienced by patients (reference category: never)			
Sometimes	0.125	0.023	
YASR Total Problems			
Sex (Male)	-0.116	0.029	0.113
Restrictions imposed by physician	0.129	0.015	
Scar judged by physician (reference category: well healed)			
Moderately healed	-0.117	0.027	
Restrictions by scar experienced by patients (reference category: never)			
Sometimes	0.224	< 0.001	
Maximum exercise capacity in %	-0.117	0.026	
YABCL Internalizing			
Course of hospitalisations as a result of heart problems Before 1990 (reference category: no hospitalisations)			
Hospitalisations without reoperation(s)	-0.121	0.028	0.059
Hospitalisations with reoperation(s)	0.122	0.026	
YABCL Externalizing			
Course of hospitalisations as a result of heart problems Before 1990 (reference category: no hospitalisations)			
Hospitalisations with reoperation(s)	0.142	0.018	0.049
Restrictions by scar experienced by patients (reference category: never)			
Sometimes	0.140	0.014	
Pacemaker	-0.158	0.009	
YABCL Total problems			
Cardiac diagnosis (reference category: ASD)			
VSD	0.121	0.034	0.069
Course of hospitalisations as a result of heart problems Before 1990 (reference category: no hospitalisations)			
Hospitalisations without reoperation(s)	-0.130	0.021	
Hospitalisations with reoperation(s)	0.147	0.010	
Restrictions by scar experienced by patients (reference category: never)			
Sometimes	0.135	0.017	

Note. Abbreviations used are YASR = Young Adult Self-Report, YABCL = Young Adult Behavior Checklist, ASD = Atrial Septal Defect, VSD = Ventricular Septal Defect, TGA = Transposition of the Great Arteries.

arteries were associated with higher scores on YASR Externalizing. Ventricular septal defect was also associated with higher scores on YABCL Total problems. Restrictions imposed by physician significantly predicted higher scores on YASR Total problems. ‘Scar judged by physician (moderately healed)', hospitalisations without reoperation(s) before 1990 and having a pace-maker were associated with lower (that is favourable) scores on some outcome variables.

R^2 is the percentage of variance in the outcome variable explained by the final predictors. Prediction variables of the YASR outcome explained higher percentages of variance than the prediction variables of the YABCL outcome on the parallel scales.

Discussion

Scar

Remarkably, the subjective prediction variable 'restrictions by scar experienced by patients' predicts the widest range of problems. The variable 'scar judged by physician' yields conflicting results. On some outcome variables, a moderately healed scar seems less predictive for psychopathology compared to a well healed scar. Further analyses showed that the extent to which patients experience restrictions by the scar and the way physicians judge the scar show little agreement ($\chi^2 = 7.98$; *df* = 4; p = 0.092). Whether a congenital heart disease patient feels restricted by the scar thus seems determined by subjective experiences and not so much by esthetical aspects. Experience of restrictions by the scar might lead to behavioural and emotional problems. This finding confirms earlier suggestions that the scar might play an important role in the well-being of adult congenital heart disease patients and can be a source of uncertainties, e.g. in sexual relationships (van Rijen et al., submitted b). Having a scar as a result of open heart surgery in itself might be a burden for congenital heart disease patients. Its inevitable presence probably is of greater importance to congenital heart disease patients than esthetical gradations. The scar reminds adult congenital heart disease patients of the open heart surgery they underwent in childhood. For some patients the scar might symbolise being different from others. The reason why congenital heart disease patients who sometimes feel restricted by the scar have a greater chance of behavioural and emotional problems than patients who often feel restricted by the scar, remains unclear.

Early hospitalisations

Reoperation before 1990 forms an important predictor in the other informant report (YABCL) of psychopathology. Remarkably, the medical course across the last decade (after 1990) does not seem to predict psychopathology in congenital heart disease patients, as reported by other informants as well as by patients themselves. An explanation could be that hospitalisations with reoperation in childhood or adolescence have a greater impact for the congenital heart disease patient than the occurrence of such medical experiences in adulthood. However, reoperations before 1990 are not of any importance in the self-report (YASR) of psychopathology. A more likely explanation thus might be that hospitalisations with reoperation performed in childhood or adolescence have a greater impact for significant others in the environment of the congenital heart disease patient than reoperations which occur in adulthood. In childhood, parents are closely involved by the care of their child and feel particularly responsible when their child is more vulnerable as a result of congenital heart disease. Parents' concerns and anxieties about the development of the child with congenital heart disease might continue into adulthood and might influence the image significant others (e.g. spouses) have of them as adults. This explanation is supported by a study of DeMaso et al. (1991) who found that maternal perceptions were more potent predictors of emotional adjustment in congenital heart disease children, compared to medical severity. It remains unclear why hospitalisations before 1990 without reoperation seem less predictive for psychopathology compared to no hospitalisations before 1990.

Sex differences

Being female significantly predicts higher rates of particularly internalising problems, e.g. being anxious or depressed. Previously, higher rates of psychopathology for female patients compared to male patients were found in this same cohort. This trend is also seen in the general population (Hofstra et al., 2000). However, especially young congenital heart disease females showed significantly more behavioural and emotional problems than a sex- and age-comparable reference group (van Rijen et al., submitted b). Being female can thus be considered as a specific predictor for psychopathology in congenital heart disease adults.

Maximum exercise capacity

Low maximum exercise capacity also predicts higher rates of particularly internalising problems. In this context, clinical observations show that, during psychological examination, many congenital heart disease patients reported to think of the bicycle ergometry as an unpleasant and fearful experience. For adult congenital heart disease patients, being in poor physical condition might lead to concerns and anxieties about one's health. Fredriksen et al. (2000) found a decrease of internalising problems in children and adolescents with congenital heart disease who were submitted to a physical training intervention. Possibly, for congenital heart disease adults, physical training might have the same favourable effect and should be considered in these patients.

Cardiac diagnosis

Patients who underwent surgery for ventricular septal defect and transposition of the great arteries have a higher risk of developing particularly externalising problems, e.g. intrusive and aggressive behaviour. Previously, differences in psychopathology found between different diagnostic categories in this same cohort were limited to higher attention problems for patients with ventricular septal defect and tetralogy of Fallot (van Rijen et al., submitted b). These latter results were found by analysis of variance in a selection of the patient sample (20-32-year-olds), whereas the present results are based on linear regression analysis for the complete patient sample with a broader age range, which might be a better method to reveal differences between diagnostic categories. It is striking that the diagnostic category transposition of the great arteries is more prominently present as significant predictor in phase 1 and 2 compared to the final model in phase 3. Since transposition of the great arteries can be considered as a severe condition with far-reaching consequences for the adult patient's life (Roos-Hesselink et al., submitted), it is probable that they also have high ratings on the other prediction variables. Correction for the other prediction variables in the final model might explain why transposition of the great arteries has receded more to background. This would imply that factors associated with the diagnosis transposition of the great arteries, such as lower maximum exercise capacity, rather than the diagnosis itself, are predictive for psychopathology at adult age. In a study of children with cyanotic heart defects, similar results were found by DeMaso et al. (1990). They stated that the diagnosis of a severe cyanotic heart defect does not appear to make a child more likely to have emotional disorder in the absence of other factors, such as impairment of the central nervous system. Higher ratings of externalising problems in patients with ventricular septal defect and transposition of the great arteries might be related to overcompensation of restrictions experienced as a result of the congenital heart disease, since risky and defiant behaviour have been reported during psychological examination.

Restrictions imposed by physician

Restrictions imposed by a physician form a significant predictor for overall problems as reported by patients themselves. This confirms earlier assumptions that experience of restrictions as a result of the congenital heart disease might hamper patients in their well-being (van Rijen et al., submitted b). Besides restrictions that are imposed by a physician, restrictions that congenital heart disease patients themselves experience subjectively, as a result of uncertainties or misconceptions, might also play a role in the occurrence of behavioural and emotional problems.

Pace-maker

Remarkably, having a pace-maker (which was the case for 30 patients in the sample) was a significant predictor for less externalising problems as reported by significant others. Alpern et al. (1989) found overall normal psychosocial adaptation of children with cardiac pace-makers. Little is known about the adjustment of adult congenital heart disease patients with pace-makers. Alpern et al. (1989) found that the self-image of young pace-makers patients is not different from that of peers, while peers assume that children and adolescents with pace-makers are more anxious, depressed, embarrassed, socially isolated, or diminished in their sense of self-worth. Similar assumptions might underlie the low externalising behaviour as reported by significant others.

Different informants

It is striking that some predictors of psychopathology in congenital heart disease adults are informant-specific. Predictors that are exclusively predictive for psychopathology as reported by the patients themselves, project the recent personal situation of the patient (maximum exercise capacity, restrictions imposed by physician). Predictors that are exclusively predictive for psychopathology as reported by significant others, mostly project the early course of hospitalisations. Previously in this same cohort, it was found that significant others reported higher ratings of psychopathology than patients themselves, which might indicate denial mechanisms in adults with congenital heart disease (van Rijen et al., submitted b). From the present results it might be concluded that, although psychopathology might be underreported by patients themselves, the level of psychopathology reported by congenital heart disease patients has a stronger relation with the recent medical status and relevant subjective experiences compared to the level of psychopathology reported by significant others. In the psychological evaluation of congenital heart disease patients, it thus seems of particular importance to take the role of different informants carefully into account.

Conclusion

In a similar prediction study, based on the first follow-up (1989-1991) of this same cohort (Utens et al., 1998b), the predictors identified mostly originated from the clusters of medical history and heart surgery. Since the earlier prediction study of this cohort of Utens et al.

(1998b) involved only children and adolescents, these predictors can be considered as relevant and topical events in their situation back then. The present study shows that early hospitalisations are still relevant in the prediction of behavioural and emotional problems as perceived by significant others, whilst recent events are more important as predictors for later problems as reported by patients themselves. Variables concerning the first open heart surgery and direct post-operative course do no longer seem to be significantly associated with the occurrence of psychopathology in adulthood. According the congenital heart disease patients' self-report, current issues as subjective experiences of the scar, physical condition and restrictions imposed by physicians are more predictive for behavioural and emotional problems. These issues could be a special area of attention in the contacts with physicians. In this context, it should be noted that objective judgements by physicians are not necessarily in accordance with subjective experiences by patients and that the latter are most likely to determine the congenital heart disease patients' well-being. It thus seems important to pay attention to the patients' report of discomfort concerning issues related to the congenital heart disease. If necessary, referral to psychological counselling should be provided. Regarding physical condition, monitored physical training might help adult congenital heart disease patients to feel more secure about their bodily functioning.

7 General discussion

Chapter 7

General discussion

Present psychosocial functioning in adult patients with congenital heart disease

In this cohort study, the psychosocial functioning of a sample of 362 adult patients with congenital heart disease was examined.

Regarding biographical characteristics, living conditions and marital status were comparable to those of the normal population. The majority of the patient sample was living independently and was in some sort of a relationship. The proportion of patients with a history of special education was high. Patients showed lower educational and occupational levels compared to reference groups. In contrast with these last findings, the patients showed normal duration of employment and a higher income compared to the normal population. Patients had a positive perception of their career possibilities. Also, the patients' social functioning, as measured by leisure-time activities, was favourable compared to reference groups.

Compared to a reference group, patients showed more favourable results on the following three scales reflecting emotional functioning: hostility, self-esteem and neuroticism. However, they showed higher total problems scores indicating psychopathology compared to a reference group. This was found for self-reports as well as other informant reports (completed by a familiar person). On the level of specific problem scales, significant higher problems scores according to the self-report were found only for the Somatic Complaints scale. The higher total problems score according to the other informant report reflected a wide range of specific syndromes. The differences in outcomes of patients between emotional functioning and psychopathology could be explained by the way items are formulated in the concerning questionnaires. Emotional functioning was assessed by more commonly formulated items concerning abstract personality traits (personal characteristics or preferences), whereas psychopathology was assessed by more concrete items concerning observable emotional and behavioural problems. Possibly, overcompensation in adult patients with congenital heart disease underlies the report of strong personality traits, while the existence of concrete emotional and behavioural problems can not easily be denied.

Lower active problem solving was found in the patient sample, which can be mainly attributed to the female patients. The patients reported to receive less social support, but also to miss social support less often compared to a reference group. Especially regarding emotional support or feedback, the patient sample seemed to feel rather independent.

Research into subjective health status revealed that patients with congenital heart disease experience more impairment on physical activities compared to a reference group. On the other hand however, the patients reported less experience of bodily pain, less interference of physical and emotional problems with normal social activities and less problems with work or other daily activities as a result of emotional problems, compared to a reference group.

In conclusion, this leads to the overall picture that adult patients with congenital heart disease, despite some residual effects concerning education, occupation and physical functioning, seem capable of leading normal lives. Their independent attitude towards social support and the low interference of physical and emotional problems with daily life seem to illustrate their strong motivation and strive of making good use of their abilities. Despite the favourable outcome on emotional functioning, further results showed elevated levels of emotional and behavioural problems in adults with congenital heart disease, particularly according to the other informant report.

Longitudinal developments in psychosocial functioning

Since this study concerned the second follow-up of a cohort study, and some of the instruments had also been used during the first follow-up, longitudinal comparisons could be made over a 10 year period. At the time of the first follow-up, the patients' ages ranged from 10 to 35 years. During the second follow-up, the age range was 20 to 46 years.

Regarding psychopathology, longitudinal comparisons were made between the first and second follow-up. Measurements from childhood, adolescence, and early adulthood (first follow-up) were compared with those of (later) adulthood, 10 years later. The comparison between early and later adulthood showed the strongest correlation. The predictive value of emotional and behavioural problems during the first follow-up for still displaying these problems 10 years later, was highest for the developmental course from early into later adulthood and, in addition, higher for males compared to females. Furthermore, longitudinal comparisons over a 10 year period showed that male patients and patients in later adulthood showed less variability in the level of problems across time compared to respectively female patients and young adult patients.

The longitudinal course of psychological well-being showed that, although feelings of displeasure increased somewhat from late adolescence/early adulthood into (later) adulthood, congenital heart disease patients became less inhibited in social situations. Male patients reported a small increase in well-being, whereas female patients reported a small decrease in well-being.

In summary, female patients with congenital heart disease seem to display more variability in the level of emotional and behavioural problems over time, compared to male patients. Furthermore, psychopathological symptoms in patients with congenital heart disease become more persistent as they grow older. This implies that assessment and treatment of psychopathology at young age is highly desirable in patients with congenital heart disease.

Medical predictors

Although the prediction variables included in the analyses represented the life time medical course, the variables which were most predictive for emotional and behavioural problems as reported by patients themselves originated from the patients' present adult lives.

The surgical scar

Restrictions by the scar as experienced by patients themselves predict the widest range of emotional and behavioural problems (internalising as well as externalising problems). The extent to which patients experience restrictions by the scar and the way physicians judge the esthetical aspects of the scar show little agreement. Furthermore, psychopathology cannot be adequately predicted from the physician's judgement of the scar. For adult patients with congenital heart disease, the inevitable presence of the scar itself might cause emotional discomfort. Whether the scar could have looked better or worse doesn't seem to matter too much. Presumably, adults with congenital heart disease don't compare themselves with each other, but rather with healthy, unoperated individuals from their own environment. The surgical scar might be a constant reminder of the open heart surgery and the congenital heart disease. Although some patients, during psychological examination, reported to be proud of their scar, 18.9% of the male patients and 40.5% of the female patients reported to feel restricted by the scar. Subjective experiences of the scar, rather than its esthetical aspects, should thus be the focus of attention when judging the patient's well-being.

Maximum exercise capacity

Low maximum exercise capacity at present is a significant predictor for particularly internalising problems. An explanation might be that poor physical condition causes fears and feelings of insecurity about one's health, especially with a medical history of a congenital heart disease.

Restrictions imposed by a physician

Restrictions imposed by a physician, as reported during the second follow-up, form a significant predictor for overall problems. Being prevented from certain aspects of normal adult life, such as participating in sports, choice of career or having offspring can have a negative influence on the patient's well-being. It is noteworthy to mention that, besides restrictions imposed by physicians, adult patients with congenital heart disease come across other limitations, for example when getting a mortgage. Moreover, patients themselves might feel or believe they are restricted by the congenital heart disease, independently of the physician's judgement. When patients no longer visit a cardiologist for medical check-ups on a regular basis, they might lack information about their medical condition. This might lead to unnecessary uncertainties or even misconceptions about one's abilities or restrictions concerning the congenital heart disease. For example, 20.1% of the male patients and 44.4% of the female patients reported that the congenital heart disease somehow was or had been a limiting factor in the choice of having children. This largely exceeds the small proportion of patients that actually have restrictions imposed by a physician.

The role of sex and age

Significantly more 25-29-year-old female patients were living on their own than same aged peers from a reference group. With respect to offspring, the patients seemed to raise families somewhat later in their twenties compared to a reference group.

Female patients showed less favourable results on neuroticism and self-esteem than male patients. This trend is in accordance with the trend in the general population. Research into psychopathology showed that, compared to a reference group, greater differences were found between males and females in the patient sample, with male patients scoring more favourably than female patients. Also, compared to the reference group, greater differences between patients in early and later adulthood were found, with patients from later adulthood scoring more favourably than those from early adulthood. For a few problems, this difference was found especially among female patients. Furthermore, being female significantly predicts higher rates of particularly internalising problems.

Research into coping styles showed favourable results for male patients and, reversely, unfavourable results for female patients. Compared to a reference group, male patients reported to seek social support (share feelings) more often, have less passive patterns of reactions and express emotions of anger and annoyance less often than a reference group. On the other hand, female patients reported to have less active problem solving, seek social support less often and show more passive patterns of reaction compared to a reference group. Although the coping styles in the female patients appeared to have unfavourable consequences for receiving social support, female patients did not experience higher discrepancies between received and desired social support. Older male patients experienced higher discrepancy between desired and received instrumental support compared to younger male patients. This might indicate a prolonged dependency on practical help, which young adults are more likely to receive than older adults.

More favourable subjective health experiences regarding physical functioning, social functioning, general mental health and vitality were found for male patients compared to female patients. These results are in line with differences between sexes found in the normal population. Remarkably, younger female patients reported more role limitations due to physical functioning compared to older female patients, whereas for male patients, the reversed situation was true.

Overall, the results indicated that especially young female patients are at risk for difficulties in psychosocial functioning. Possible problems young female patients might experience are described below.

Young female patients at risk

Although some unfavourable outcomes for female patients (e.g. emotional functioning and subjective health experiences) are in accordance with patterns found in the normal population, the unfavourable results on psychopathology and coping styles show their disadvantageous position compared to reference peers.

There are some clear indications that especially young female patients are at risk for experiencing problems. Firstly, compared to a reference group, larger differences in emotional and behavioural problems between younger and older female patients were found, with the younger female patients scoring less favourably than the older female patients. Secondly, younger female patients seem to feel more limited due to their physical functioning compared to older female patients. These findings indicate that young female patients struggle with certain aspects of life that are apparently not so prominent in later adulthood.

Research into prediction variables provides some insight in what these issues might be. Firstly, there is the surgical scar, which is unfavourably situated on the chest. Especially for young females, the scar can therefore be a source of uncertainties or discomfort, for example when entering sexual relationships. Secondly, uncertainties regarding abilities of pregnancy and delivery might play a role. For female patients, these issues concern their own bodily functioning and might therefore cause anxiety about their physical condition. The high proportion of especially female patients who reported to feel or had felt restricted somehow by the congenital heart disease in their choice of having children (see “restrictions imposed by a physician”), might explain why the patient sample seemed to raise families somewhat later compared to the normal population. The assumption that the issues of relations and offspring strike young female patients in particular, is supported by the theoretical model of Cyranowski et al. (2000). According to Cyranowski et al. (2000), the reason why young females are more likely than young males to become depressed when faced with negative life events is due to a preference of females for close emotional communication, intimacy and responsiveness within interpersonal relationships. Therefore, negative consequences of the congenital heart disease on life events which are prominent in early adulthood, such as relations and offspring, are likely to strike females harder than males. Moreover, female patients, unlike male patients, don’t seem to have very adequate coping styles at their disposal. Their less active problem solving and more passive patterns of reaction might reflect feelings of having no control over the problems they encounter. In summary, when facing obstacles associated with congenital heart disease in adulthood, young females display larger vulnerability to these problems and less adequate coping styles. Presumably, these disease-related problems and uncertainties recede more to the background once a stable relationship and/or family life are established. This might also explain the greater difference in emotional and behavioural problems between early and later adulthood in patients with congenital heart disease, compared to a reference group.

The role of the cardiac diagnosis

In comparison to other cardiac diagnostic categories, patients from the diagnostic categories tetralogy of Fallot and transposition of the great arteries, showed trends to unfavourable results regarding former course of education, educational level, social security benefits, duration of employment and sick leave. However, statistical significant differences could only be revealed regarding the former course of education. Patients with transposition of the great arteries showed lower participation in intensive sport.

No statistical differences for emotional functioning were found between cardiac diagnostic categories. With respect to psychopathology, higher rates of attention problems were found in patients with ventricular septal defect and tetralogy of Fallot. Research into prediction variables showed that patients with ventricular septal defect and transposition of the great arteries have a higher risk of developing particularly externalising problems. The latter results might reflect overcompensation, since risky and defiant behaviour have been reported.

In contrast to the other diagnostic categories, older patients with tetralogy of Fallot and pulmonary stenosis showed trends to more passive patterns of reaction compared to younger patients within the same cardiac diagnostic categories. No further differences between cardiac diagnostic categories were found for either coping styles or social support.

Although no significant differences between diagnostic categories were revealed concerning subjective health experiences, there were some indications that patients with transposition of the great vessels do experience some disadvantageous consequences of the congenital heart disease on their physical health. In accordance with previous studies (Greenwood, 2000; Kamphuis et al., 2002) this diagnostic category showed the poorest outcomes as to physical functioning, vitality and general health perceptions. In contrast, patients with transposition of the great arteries, along with patients with tetralogy of Fallot, obtained by far the most favourable scores for experience of bodily pain.

In summary, for patients with tetralogy of Fallot and transposition to the great arteries, a slight trend in the negative direction was found. To a lesser extent, there were a few unfavourable findings for patients with ventricular septal defect and pulmonary stenosis. Patients with atrial septal defect overall showed favourable psychosocial functioning, compared to other diagnostic groups. These outcomes seem in line with the medical severity of the cardiac diagnoses. Patients with tetralogy of Fallot and transposition of the great arteries can be considered as most severely affected. Patients with ventricular septal defect and pulmonary stenosis may still experience residual effects from the congenital heart disease, and to a much lesser extent this is the case for patients with atrial septal defect. Considering the large number of aspects of psychosocial function that were assessed however, these slight trends for different cardiac diagnoses were only observed in a scanty number of variables. Possible explanations are given below.

Explanations for lack of significant differences between cardiac diagnostic groups

For most aspects of psychosocial functioning, no significant differences between diagnostic categories could be revealed. A possible explanation why significant differences between diagnostic categories could only scarcely be found are the relative large confidence intervals for the separate diagnostic groups, due to smaller cells, compared to the complete patient sample. Our findings concerning prediction variables offer a second explanation, namely that factors associated with the cardiac diagnosis, such as exercise capacity, rather than the diagnosis itself are responsible for poor functioning. Thirdly, in accordance with clinical observations during the psychological examination, we might hypothesise that patients from different diagnostic categories apply different approaches to disease when judging the medical severity of their congenital heart disease. Patients who actually are impaired and restricted by the congenital heart disease might choose to fight and deny these negative consequences in order to live a normal life as much as possible. This is well illustrated by the low report of bodily pain in patients with tetralogy of Fallot and transposition of the great arteries. On the other hand, patients whose medical condition no longer requires medical check-ups by a cardiologist, might lack the information and attention for their congenital heart disease (Ferencz et al., 1980; Moons et al., 2001), which might lead to fears and uncertainties. Patients then, might create their own interpretation of the severity of the congenital heart disease and may thereby be oversensitive to or overestimate the negative consequences of the congenital heart disease. This might be especially true for this cohort, since these patients were the first to receive open heart surgery for congenital heart disease in this hospital. The tension and excitement about open heart surgery on children back then, might be very much in contrast with the relatively calm attitude towards mild congenital heart diseases that patients perceive today. Processes of both under- and overestimation of the medical severity of the congenital heart disease might thus occur in adults with congenital heart disease. This might explain why differences in psychosocial functioning between different cardiac diagnostic groups are limited.

Different informants

For psychopathology, a differential informant effect was found, indicating a greater difference between self and other informant report in the patient sample compared to the reference group. Other informants seemed to evaluate psychopathological symptoms in adult congenital heart disease patients as more deviant from the norm than patients themselves. This might indicate denial mechanisms in adults with congenital heart disease. Research into prediction variables revealed that, whereas psychopathology as reported by patients themselves could best be predicted from variables of their present adult life (e.g. physical condition, restrictions), predictors for psychopathology as reported by significant others mostly originated from the early medical course (e.g. hospitalisations with reoperations before 1990). This might indicate that the medical course in childhood and adolescence still influences how significant others evaluate the patient’s functioning at adult age. Although adults with congenital heart disease might thus have a tendency to underreport problems, their self-report of psychosocial functioning shows a stronger relation with the recent medical status and relevant subjective experiences compared to the other informant report.

Strengths and limitations

The present study encompasses a consecutive series of five large diagnostic categories. A total of 362 patients participated in this study. The overall response rate was 90.7%. Various aspects of psychosocial functioning were addressed, using (semi-) structured and standardised methods. Since this study concerned the second follow-up of this cohort, longitudinal comparisons over the past 10 years were made for indicators of psychosocial functioning. Also, medical predictors, originating from the complete medical course from birth until the present, on long-term emotional and behavioural problems were identified. Overall, these results provided information regarding psychosocial functioning in adults with congenital heart disease, about which little was known so far.

Besides these profits, the study has some limitations. The patient sample contained a selection of five frequently occurring diagnostic categories in patients from only one university hospital and may therefore not be completely representative of all patients with congenital heart anomalies. The patient sample was treated with different techniques than used at present. The results may therefore not (fully) apply to the present generation of children operated for congenital heart disease nowadays. This study did not include a randomly chosen control group of healthy peers, which was matched for age and sex. Since suitable normative data were available for most instruments, comparisons could be made between the patient sample and reference groups. A part of the present patient sample was still in childhood or adolescence at the time of the first follow-up. Longitudinal comparisons from early into later adulthood were therefore only available for a selection of the patient sample. Only a few instruments had parallel versions for childhood, adolescence and adulthood, enabling longitudinal comparisons from childhood and adolescence into adulthood.

Clinical implications

Overall, psychosocial functioning in adults with congenital heart disease was satisfactory. Besides favourable outcomes however, this study identified several obstacles and weaknesses in the psychosocial functioning of adults with congenital heart disease. These insights can be applied to meet the needs of this patient population more adequately. Therefore, the following clinical implications are given.

1. Assessment and treatment of emotional and behavioural problems in patients with congenital heart disease should be provided at an early stage. Although psychopathological symptoms seem to diminish in the transition form early into later adulthood, at young age the symptoms seem less persistent and thus better treatable. Moreover, when treating the patient's emotional and behavioural problems and possible underlying disease-specific uncertainties, the struggle into adulthood might be less hard, and therefore improve the patient's quality of life.
2. Especially young female congenital heart disease patients are at risk for poor psychosocial functioning. They seem more vulnerable to obstacles associated with congenital heart disease in adulthood and have less adequate coping styles at their disposal compared to male patients. Special attention should be paid to restrictions they experience subjectively regarding the scar and abilities of pregnancy and delivery. Providing better information on the medical condition might relief the patient's uncertainties. Psychological intervention, for example behavioural counselling aimed at replacing passive reaction patterns by active problem solving, might help female patients to feel more in control of their situation.
3. When assessing the adult congenital heart disease patient's well-being, subjective experiences regarding the surgical scar, rather than its esthetical aspects, should be evaluated.
4. Monitored physical training might help congenital heart disease patients with poor physical condition to feel more secure about their bodily functioning.
5. When imposing restrictions to adult congenital heart disease patients, the physician should not only discuss the patient's compliance, but also the patient's personal experiences of psychological burden following the restrictions.
6. In psychological examination of adults with congenital heart disease, the role of different informants should be taken into account. Although denial mechanisms and overcompensation might result in underreporting of emotional and behavioural problems by patients themselves, their self-report is strongly associated with their recent medical course and experiences. Regarding the other informants' report (e.d. spouses, parents), early hospital experiences seem to play a larger role in reporting emotional and behavioural problems in the patient.
7. Family doctors or general physicians should be well-informed about the medical course and consequences of congenital heart disease in adulthood, so that adults with congenital heart disease can have wide and easy access to accurate information regarding their health condition.

8. Cardiologists should have the opportunity to refer congenital heart disease patients with psychosocial problems to psychologists who are specialised is this field.

Conclusion

Besides some residual effects (especially for patients with tetralogy of Fallot and transposition of the great arteries) this cohort of adult patients with congenital heart disease overall seemed capable of leading normal lives. They seemed motivated to make good use of their abilities. Disease-related uncertainties and (subjective) experiences of restrictions as a result of the congenital heart disease might lead to elevated levels of emotional and behavioural problems. Especially (young) female patients seem to be at risk for developing psychopathology. Providing accurate information about the patient’s medical status and early assessment and treatment of emotional and behavioural problems are desirable in adults with congenital heart disease.

References

References

Aaronson, N. K., Muller, M., Cohen, P. D. A., Essink-Bot, M. L., Fekkes, M., Sanderman, R., Sprangers, M. A.G., te Velde, A., & Verrips, E. (1998). Translation, validation, and norming of the Dutch language version of the SF-36 Health Survey in community and chronic disease populations. *Journal of Clinical Epidemiology, 51*(11), 1055-1068.

Achenbach, T. M. (1990). *The Young Adult Self-Report.* Burlington: University of Vermont, Department of Psychiatry.

Achenbach, T. M. (1991a). *Manual for the Child Behavior Checklist/4-18 and 1991 Profile.* Burlington: University of Vermont, Department of Psychiatry.

Achenbach, T. M. (1991b). *Manual for the Youth Self-Report and 1991 Profile.* Burlington: University of Vermont, Department of Psychiatry.

Achenbach, T. M. (1997). *Manual for the Young Adult Self-Report and Young Adult Behavior Checklist.* Burlington: University of Vermont, Department of Psychiatry.

Alden, B., Gilljam, T., & Gillberg, C. (1998). Long-term psychological outcome of children after surgery for transposition of the great arteries. *Acta Paediatrica, 87*(4), 405-410.

Allen, H. D., Gersony, W. M., & Taubert, K. A. (1992). Insurability of the adolescent and young adult with heart disease. Report from the Fifth Conference on Insurability, October 3-4, 1991, Columbus, Ohio. *Circulation, 86*(2), 703-710.

Alpern, D., Uzark, K., & Dick, M., 2nd (1989). Psychosocial responses of children to cardiac pacemakers. *The Journal of Pediatrics, 114*(3), 494-501.

Baer, P. E., Freedman, D. A., & Garson, A. (1984). Long-term psychological follow-up of patients after corrective surgery for tetralogy of Fallot. *Journal of the American Academy of Child Psychiatry, 23*(5), 622-625.

Bellinger, D. C., Rappaport, L. A., Wypij, D., Wernovsky, G., & Newburger J. W. (1997). Patterns of developmental dysfunction after surgery during infancy to correct transposition of the great arteries. *Journal of Developmental and Behavioral Pediatrics, 18*(2), 75-83.

Bellinger, D. C., Wypij, D., Kuban, K. C. K., Rappaport, L. A., Hickey, P. R., Wernovsky, G., Jonas, R. A., & Newburger, J. W. (1999). Developmental and neurological status of children at 4 years of age after heart surgery with hypothermic circulatory arrest or low-flow cardiopulmonary bypass. *Circulation, 100*(5), 526-532.

Brandhagen, D. J., Feldt, R. H., & Williams, D. E. (1991). Long-term psychologic implications of congenital heart disease: a 25-year follow-up. *Mayo Clinic Proceedings, 66*(5), 474-479.

Carey, L. K., Nicholson, B. C., & Fox, R. A. (2002). Maternal factors related to parenting young children with congenital heart disease. *Journal of Pediatric Nursing, 17*(3), 174-183.

Casey, F. A., Craig, B. G., & Mulholland, H. C. (1994). Quality of life in surgically palliated complex congenital heart disease. *Archives of Disease in Childhood, 70*(5), 382-386.

Celermajer, D. S., & Deanfield, J. E. (1993). Employment and insurance for young adults with congenital heart disease. *British Heart Journal, 69*(6), 539-543.

Cohen, J. B. (1988). *Statistical Power Analysis for the Behavioral Sciences (Second edition ed.).* Hillsdale: Lawrence Erlbaum Associates.

Cox, D., Lewis, G., Stuart, G., & Murphy, K. (2002). A cross-sectional study of the prevalence of psychopathology in adults with congenital heart disease. *Journal of Psychosomatic Research, 52*(2), 65-68.

Cyranowski, J. M., Frank, E., Young, E., & Shear, M. K. (2000). Adolescent onset of the gender difference in lifetime rates of major depression: a theoretical model. *Archives of General Psychiatry, 57*(1), 21-27.

Davis, C. C., Brown, R. T., Bakeman, R., & Campbell, R. (1998). Psychological adaptation and adjustment of mothers of children with congenital heart disease: Stress, coping, and family functioning. *Journal of Pediatric Psychology, 23*(4), 219-228.

DeMaso, D. R., Beardslee, W. R., Silbert, A. R., & Fyler, D. C. (1990). Psychological functioning in children with cyanotic heart defects. *Journal of Developmental and Behavioral Pediatrics, 11*(6), 289-294.

DeMaso, D. R., Campis, L. K., Wypij, D., Bertram, S., Lipshitz, M., & Freed, M. (1991). The impact of maternal perceptions and medical severity on the adjustment of children with congenital heart disease. *Journal of Pediatric Psychology, 16*(2), 137-149.

Ellerbeck, K. A., Smith, M. L., Holden, E. W., McMenamin, S. C., Badawi, M. A., Brenner, J. I., Kan, J. S., & Hyman, S. L. (1998). Neurodevelopmental outcomes in children surviving d-transposition of the great arteries. *Journal of Developmental and Behavioural Pediatrics, 19*(5), 335-341.

Erdman, R. A. M. (1982). *Manual for the Heart Patient Psychological Questionnaire. Handleiding bij de Medisch Psychologische Vragenlijst voor Hartpatienten.* Swets & Zeitlinger B.V.

Fekkes, M., Kamphuis, R. P., Ottenkamp, J., Verrips, E., Vogels, T. G., Kamphuis, M., & Verloove-Vanhorick, S. P. (2001). Health-related quality of life in young adults with minor congenital heart disease. *Psychology and Health, 16*, 239-250.

Ferdinand, R. F., Verhulst, F. C., & Wiznitzer, M. (1995). Continuity and change of self-reported problem behaviors from adolescence into young adulthood. *Journal of American Academy of Child and Adolescent Psychiatry, 34*(5), 680-690.

Ferencz, C., Wiegmann, F. L., Jr., & Dunning, R. E. (1980). Medical knowledge of young persons with heart disease. *The Journal of School Health, 50*(3), 133-136.

Fredriksen, P. M., Kahrs, N., Blaasvaer, S., Sigurdsen, E., Gundersen, O., Roeksund, O., Norgaand, G., Vik, J. T., Soerbye, O., Ingjer, E., & Thaulow, E. (2000). Effect of physical training in children and adolescents with congenital heart disease. *Cardiology in the Young, 10*(2), 107-114.

Gantt, L. T. (1992). Growing up heartsick: The experiences of young women with congenital heart disease. *Health Care for Women International, 13*(3), 241-248.

Gardner, F. V., Freeman, N. H., Black, A. M. S., & Angelini, G. D. (1996). Disturbed mother-infant interaction in association with congenital heart disease. *Heart, 76*(1), 56-59.

Garson, A. Jr., Benson, R. S., Ivler, L., & Patton, C. (1978). Parental reactions to children with congenital heart disease. *Child Psychiatry & Human Development, 9*(2), 86-94.

Gersony, W. M., Hayes, C. J., Driscoll, D. J., Keane, J. F., Kidd, L., O'Fallon, W. M., Pieroni, D. R., Wolfe, R. R., & Weidman, W. H. (1993). Second natural history study of congenital heart defects. Quality of life of patients with aortic stenosis, pulmonary stenosis, or ventricular septal defect. *Circulation, 87*(2 Suppl), I52-65.

Gough, H. (1964). *Manual for the California Psychological Inventory.* Palo Alto: Consulting Psychologists Press.

Greenwood, R. (2000). Living with heart: A study of health-related quality of life in adults with congenital heart disease. *Dissertation Abstracts International: Section B: the Sciences & Engineering, 60*(12-B), 6020.

Gupta, S., Giuffre, R. M., Crawford, S., & Waters, J. (1998). Covert fears, anxiety and depression in congenital heart disease. *Cardiology in the Young, 8*(4), 491-499.

Hart, E. M., & Garson, A., Jr. (1993). Psychosocial concerns of adults with congenital heart disease. Employability and insurability. *Cardiology Clinics, 11*(4), 711-715.

Hellstedt, L. F. (1994). Insurability issues facing the adolescent and adult with congenital heart disease. *Nursing Clinics of North America, 29*(2), 331-343.

Hofstra, M. B., Van der Ende, J., & Verhulst, F. C. (2000). Continuity and change of psychopathology from childhood into adulthood: a 14-year follow-up study. *Journal of the American Academy of Child and Adolescent Psychiatry, 39*(7), 850-858.

Kamphuis, M., Ottenkamp, J., Vliegen, H. W., Vogels, T., Zwinderman, K. H., Kamphuis, R. P., & Verloove-Vanhorick, S. P. (2002). Health related quality of life and health status in adult survivors with previously operated complex congenital heart disease. *Heart, 87*(4), 356-362.

Kokkonen, J., & Paavilainen, T. (1992). Social adaptation of young adults with congenital heart disease. *International Journal of Cardiology, 36*(1), 23-29.

Kramer, H. H., Awiszus, D., Sterzel, U., Van Halteren, A., & Claßen, R. (1989). Development of personality and intelligence in children with congenital heart disease. *Journal of Child Psychology and Psychiatry and Allied Disciplines, 30*(2), 299-308.

Lane, D. A., Lip, G. Y. H., & Millane, T. A. (2002). Quality of life in adults with congenital heart disease. *Heart, 88*(1), 71-75.

Lawoko, S., & Soares, J. J. F. (2002). Distress and hopelessness among parents of children with congenital heart disease, parents of children with other diseases, and parents of healthy children. *Journal of Psychosomatic Research, 52*(4), 193-208.

Linde, L. M., Rasof, B., Dunn, O. J., & Rabb, E. (1966). Attitudinal factors in congenital heart disease. *Pediatrics, 38*(1), 92-101.

Luteijn, F., Starren, J., & van Dijk, H. (1975). *Manual for the Dutch Personality Questionnaire. Handleiding bij de NPV.* Lisse: Swets & Zeitlinger B.V.

Luteijn, F., Starren, J., & van Dijk, H. (1985). *Manual for the Dutch Personality Questionnaire, revised version. Handleiding bij de NPV, herziene uitgave.* Lisse: Swets & Zeitlinger B.V.

Mahoney, L. T., Truesdell, S. C., Hamburgen, M., & Skorton, D. J. (1991). Insurability, employability, and psychosocial considerations. In J. K. Perloff & J. S. Child (Eds.), *Congenital heart disease in adults* (pp. 178-189): W.B. Saunders Company.

Masi, G., & Brovedani, P. (1999). Adolescents with congenital heart disease: psychopathological implications. *Adolescence, 34*(133), 185-191.

McGrath, K. A., & Truesdell, S. C. (1994). Employability and career counseling for adolescents and adults with congenital heart disease. *Nursing Clinics of North America, 29*(2), 319-330.

Meijboom, F., Hess, J., Szatmari, A., Utens, E. M. W. J., McGhie, J., Deckers, J. W., Roelandt, J. R., & Bos, E. (1993). Long-term follow-up (9 to 20 years) after surgical closure of atrial septal defect at a young age. *American Journal of Cardiology, 72*(18), 1431-1434.

Meijboom, F., Szatmari, A., Deckers, J. W., Utens, E. M. W. J., Roelandt, J. R. T. C., Bos, E., & Hess, J. (1995). Cardiac status and health-related quality of life in the long term after surgical repair of tetralogy of Fallot in infancy and childhood. *Journal of Thoracic and Cardiovascular Surgery, 110*(4 Pt 1), 883-891.

Meijboom, F., Szatmari, A., Deckers, J. W., Utens, E. M. W. J., Roelandt, J. R. T. C., Bos, E., & Hess, J. (1996). Long-term follow-up (10 to 17 years) after Mustard repair for transposition of the great arteries. *Journal of Thoracic and Cardiovascular Surgery, 111*(6), 1158-1168.

Meijboom, F., Szatmari, A., Utens, E., Deckers, J. W., Roelandt, J. R. T. C., Bos, E., & Hess, J. (1994). Long-term follow-up after surgical closure of ventricular septal defect in infancy and childhood. *Journal of the American College of Cardiology, 24*(5), 1358-1364.

Moons, P., De Volder, E., Budts, W., De Geest, S., Elen, J., Waeytens, K., & Gewillig, M. (2001). What do adult patients with congenital heart disease know about their disease, treatment, and prevention of complications? A call for structured patient education. *Heart, 86*(1), 74-80.

Netherlands Central Bureau of Statistics. (1986). *Continuous Survey Quality of Life 1986.* Voorburg/Heerlen: Netherlands Central Bureau of Statistics.

Netherlands Central Bureau of Statistics. (2000). *Permanent Survey Living Situation (POLS).* Voorburg/ Heerlen: Netherlands Central Bureau of Statistics.

Netherlands Central Bureau of Statistics. (2002). Voorburg/Heerlen: Netherlands Central Bureau of Statistics.

Patton, D. E., Lee, W., Cotton, D. B., Miller, J., Carpenter, R. J., Jr., Huhta, J., & Hankins, G. (1990). Cyanotic maternal heart disease in pregnancy. *Obstetrical and Gynecological Survey, 45*(9), 594-600.

Rietveld, S., Mulder, B. J. M., van Beest, I., Lubbers, W., Prins, P. J. M., Vioen, S., Bennebroek-Evererz, F., Vos, A., Casteelen, G., & Karsdorp, P. (2002). Negative thoughts in adults with congenital heart disease. *International Journal of Cardiology, 86*(1), 19-26.

Roos-Hesselink, J. W., Meijboom, F. J., Spitaels, S. E. C., van Domburg, R., van Rijen, E. H. M., Utens, E. M. W. J., Bogers, A. J. J. C., & Simoons, M. L. (2003a). Excellent survival and low incidence of arrhythmias, stroke and heart failure long-term after surgical ASD closure at young age. A prospective follow-up study of 21-33 years. *European Heart Journal, 24*(2), 190-197.

Roos-Hesselink, J. W., Meijboom, F. J., Spitaels, S. E. C., van Domburg, R. T., van Rijen, E. H. M., Utens, E. M. W. J., McGhie, J., Bogers, A. J. J. C., & Simoons, M. L. (submitted). Decline in ventricular function and clinical condition after Mustard repair for transposition of the great arteries (a prospective follow-up study of 22-29 years).

Saliba, Z., Butera, G., Bonnet, D., Bonhoeffer, P., Villain, E., Kachaner, J., Sidi, D., & Iserin, L. (2001). Quality of life and perceived health status in surviving adults with univentricular heart. *Heart, 86*(1), 69-73.

Salzer-Muhar, U., Herle, M., Floquet, P., Freilinger, M., Greber-Platzer, S., Haller, A., Leixnering, W., Marx, M., Wurst, E., & Schlemmer, M. (2002). Self-concept in male and female adolescents with congenital heart disease. *Clinical Pediatrics (Phila), 41*(1), 17-24.

Schmidt, S., Nachtigall, C., Wuethrich-Martone, O., & Strauss, B. (2002). Attachment and coping with chronic disease. *Journal of Psychosomatic Research, 53*(3), 763-773.

Schreurs, P. J. G., van de Willige, G., Brosschot, J. F., Tellegen, B., & Graus, G. M. H. (1993). *De Utrechtse Coping Lijst: UCL, omgaan met problemen en gebeurtenissen, herziene handleiding 1993.* Lisse, : Swets & Zeitlinger.

Shampaine, E. L., Nadelman, L., Rosenthal, A., Behrendt, D., & Sloan, H. (1990). Longitudinal psychological assessment in tetralogy of Fallot. *Pediatric Cardiology, 10*(3), 135-140.

Somerville, J. (1998). The Denolin Lecture: The woman with congenital heart disease. *European Heart Journal, 19*(12), 1766-1775.

Sparacino, P. S. A. (1994). Adult congenital heart disease. An emerging population. *Nursing Clinics of North America, 29*(2), 213-219.

Spurkland, I., Bjornstad, P. G., Lindberg, H., & Seem, E. (1993). Mental health and psychosocial functioning in adolescents with congenital heart disease. A comparison between adolescents born with severe heart defect and atrial septal defect. *Acta Paediatrica, 82*(1), 71-76.

Stinson, J., & McKeever, P. (1995). Mothers' information needs related to caring for infants at home following cardiac surgery. *Journal of Pediatric Nursing, 10*(1), 48-57.

Tak, Y. R., & McCubbin, M. (2002). Family stress, perceived social support and coping following the diagnosis of a child's congenital heart disease. *Journal of Advanced Nursing, 39*(2), 190-198.

Ternestedt, B. M., Wall, K., Oddsson, H., Riesenfeld, T., Groth, I., & Schollin, J. (2001). Quality of life 20 and 30 years after surgery in patients operated on for tetralogy of Fallot and for atrial septal defect. *Pediatric Cardiology, 22*(2), 128-132.

Tong, E., & Sparacino, P. S. A. (1994). Special management issues for adolescents and young adults with congenital heart disease. *Critical Care Nursing Clinics of North America, 6*(1), 199-214.

Utens, E. M., Versluis-Den Bieman, H. J., Verhulst, F. C., Meijboom, F. J., Erdman, R. A., & Hess, J. (1998a). Psychopathology in young adults with congenital heart disease. Follow-up results. *European Heart Journal, 19*(4), 647-651.

Utens, E. M., Verhulst, F. C., Duivenvoorden, H. J., Meijboom, F. J., Erdman, R. A., & Hess, J. (1998b). Prediction of behavioural and emotional problems in children and adolescents with operated congenital heart disease. *European Heart Journal, 19*(5), 801-807.

Utens, E. M., Versluis-Den Bieman, H. J., Verhulst, F. C., Witsenburg, M., Bogers, A. J. J. C., & Hess, J. (2000a). Psychological distress and styles of coping in parents of children awaiting elective cardiac surgery. *Cardiology in the Young, 10*(3), 239-244.

Utens, E. M., Versluis-Den Bieman, H. J., Witsenburg, M., Bogers, A. J. J. C., Hess, J., & Verhulst, F. C. (2002). Does age at the time of elective cardiac surgery or catheter intervention in children influence the longitudinal development of psychological distress and styles of coping of parents? *Cardiology in the Young, 12,* 524-530.

Utens, E. M. W. J. (1992). Psychosocial aspects of congenital heart disease in children, adolescents and adults. In P. J. Walter (Ed.), *Quality of Life After Open Heart Surgery* (pp. 325-331). Printed in the Netherlands: Kluwer Academic Publishers.

Utens, E. M. W. J., van Rijen, E. H. M., Erdman, R. A. M., & Verhulst, F. C. (2000b). Rotterdams Kwaliteit van Leven - Interview.

Utens, E. M. W. J., Verhulst, F. C., Erdman, R. A. M., Meijboom, F. J., Duivenvoorden, H. J., & Hess, J. (1996). Psychosociaal functioneren van jeugdigen en jong-volwassenen 9 jaar en langer na operatieve behandeling van een aangeboren hartafwijking op de kinderleeftijd. *Nederlands Tijdschrift voor Geneeskunde, 140*(21), 1126-1130.

Utens, E. M. W. J., Verhulst, F. C., Erdman, R. A. M., Meijboom, F. J., Duivenvoorden, H. J., Bos, E., Roelandt J. R. T. C. & Hess, J. (1994). Psychosocial functioning of young adults after surgical correction for congenital heart disease in childhood: A follow-up study. *Journal of Psychosomatic Research, 38*(7), 745-758.

Utens, E. M. W. J., Verhulst, F. C., Meijboom, F. J., Duivenvoorden, H. J., Erdman, R. A. M., Bos, E., Roelandt, J. T. C. & Hess, J. (1993). Behavioural and emotional problems in children and adolescents with congenital heart disease. *Psychological Medicine, 23*(2), 415-424.

van Rijen, E. H. M., Utens, E. M. W. J., Roos-Hesselink, J. W., Meijboom, F. J., van Domburg, R. T., Roelandt, J. R. T. C., Bogers, A. J. J. C., & Verhulst, F. C. (2003). Psychosocial functioning of the adult with congenital heart disease: a 20-33 years follow-up. *European Heart Journal, 24*(7), 673-683.

van Rijen, E. H. M., Utens, E. M. W. J., Roos-Hesselink, J. W., Meijboom, F. J., van Domburg, R. T., Roelandt, J. R. T. C., Bogers, A. J. J. C., & Verhulst, F. C. (submitted a). Coping styles and social support in a cohort of adults with congenital heart disease.

van Rijen, E. H. M., Utens, E. M. W. J., Roos-Hesselink, J. W., Meijboom, F. J., Van Domburg, R. T., Roelandt, J. R. T. C., Bogers, A. J. J. C., & Verhulst, F. C. (submitted b). Development of psychopathology over a 10 year period in a cohort of adults with congenital heart disease.

van Sonderen, E. (1993). *Het meten van sociale steun met de Sociale Steun Lijst - Interacties (SSL-I) en Sociale Steun Lijst - Discrepanties (SSL-D), een handleiding.* Groningen: Noordelijk Centrum voor Gezondheidsvraagstukken.

Verhulst, F. C., van der Ende, J., & Koot, H. M. (1996). *Handleiding voor de CBCL/4-18 [Manual for the CBCL/4-18].* Rotterdam: Erasmus University/Department of Child and Adolescent Psychiatry, Sophia Children's Hospital.

Verhulst, F. C., van der Ende, J., & Koot, H. M. (1997). *Handleiding voor de Youth Self-Report (YSR) [Manual for the Youth Self-Report (YSR)].* Rotterdam: Erasmus University/Department of Child and Adolescent Psychiatry, Sophia Children's Hospital.

Visconti, K. J., Saudino, K. J., Rappaport, L. A., Newburger, J. W., & Bellinger, D. C. (2002). Influence of parental stress and social support on the behavioral adjustment of children with transposition of the great arteries. *Journal of Developmental and Behavioral Pediatrics, 23*(5), 314-321.

Ware, J. E., Jr., & Sherbourne, C. D. (1992). The MOS 36-item short-form health survey (SF-36). I. Conceptual framework and item selection. *Medical Care, 30*(6), 473-483.

World Health Organisation. (1948*). Constitution of the World Health Organization basic documents.* Geneva: World Health Organization.

Wray, J., & Sensky, T. (2001). Congenital heart disease and cardiac surgery in childhood: effects on cognitive function and academic ability. *Heart, 85*(6), 687-691.

Wren, C., & O'Sullivan, J. J. (2001). Survival with congenital heart disease and need for follow up in adult life. *Heart, 85*(4), 438-443.

Wright, M., Jarvis, S., Wannamaker, E., & Cook, D. (1985). Congenital heart disease: functional abilities in young adults. *Archives of Physical and Medical Rehabilitation, 66*(5), 289-293.

Summary

Summary

The objective of the present study was to examine a broad range of psychosocial aspects of congenital heart disease in adulthood. In **chapter 1**, the background of the current study was presented. Since over the last decades, as a result of improvements in medical treatment, diagnostic and surgical techniques, mortality rates for congenital heart disease decreased, more patients have survived into adulthood. Adults with congenital heart disease form a new patient population. Little is yet known about their psychosocial functioning. In the present cohort study, various aspects of psychosocial functioning were examined during psychological examination in 362 adults with congenital heart disease, 20-33 years after their first open heart surgery. Since this study was the second follow-up of the cohort, we were able to examine the patients' present functioning, as well as the longitudinal development over the past 10 years. The main aims of the study were: (1) to compare the present psychosocial functioning of adults with congenital heart disease with that of normative samples, (2) to determine the 10 year longitudinal course of psychosocial functioning of adults with congenital heart disease, (3) to determine the role of age, gender and cardiac diagnostic category on psychosocial functioning of adults with congenital heart disease, (4) to determine the predictive value of a wide range of medical variables on emotional and behavioural problems of adults with congenital heart disease.

In **chapter 2**, biographical characteristics (e.g. living conditions, offspring, education, marital and occupational status), emotional functioning (the scales Hostility, Self-esteem and Neuroticism of the Dutch Personality Questionnaire) and social functioning (leisure-time activities) were examined. Living conditions and marital status were comparable to those in the normal population. Rates of offspring appeared favourable, though none of the 20-24-year-old married or cohabitant patients had offspring. The patient sample showed lower educational and occupational levels compared to the normal population, but otherwise showed favourable participation in employment. The patient sample showed favourable emotional functioning. They reported less hostility and neuroticism and better self-esteem compared to a reference group. Compared to the normal population, the patients' leisure-time activities were favourable. Overall, the patients seemed capable of leading normal lives and seemed motivated to make good use of their abilities.

In **chapter 3**, psychopathology (emotional and behavioural problems) in the patient sample was examined currently as well as longitudinally over a 10 year period. In the present

study patients filled in the Young Adult Self-Report and another informant (e.g. parent, spouse) filled in the Young Adult Behavior Checklist about the patient. Ten years before, the Young Adult Self-Report was also administered. For patients who then had not yet reached adulthood, parallel versions of these questionnaires were used for childhood and adolescence (Child Behavior Checklist and Youth Self-Report). Young female patients showed higher levels of psychopathology compared to peers from the normal population. Parents and partners of patients evaluated the patients' behaviour as more deviant from the norm than patients themselves. Younger patients showed more psychopathology than older patients. Also, female patients showed more psychopathology than male patients. Furthermore, longitudinal comparisons over a 10 year period showed that younger and female patients showed more variability (increase or decrease) in the level of problems across time compared to older and male patients respectively. The results show the need for assessment and treatment of psychopathology at young age. Psychopathology in young female patients, and possibly underlying disease-specific uncertainties concerning relationships and offspring, also deserve special attention.

In **chapter 4**, coping styles (personal ways of reacting to problems) and social support (support from the social environment) in the patient sample were assessed with the Utrecht Coping List and Social Support List respectively. Female patients showed less favourable coping styles compared to reference females, such as less active problem solving. Male patients showed more favourable coping styles compared to reference males, such as higher seeking of social support, lower passive reaction patterns and lower expression of (negative) emotions. Compared to the reference group, the total patient sample reported to receive less social support, but also to experience less discrepancies between desired and received social support, indicating feelings of independence in these adult patients. Results indicated that the less active problem solving in females with congenital heart disease needs special attention.

In **chapter 5**, the present subjective health status and the longitudinal course of psychological well-being over a 10 year period in the patient sample were examined by the SF-36 and Heart Patients Psychological Questionnaire respectively. The patient sample reported less favourable subjective health status concerning physical functioning compared to a reference group. On the contrary, the patient sample reported better social functioning, less bodily pain and less role limitations due to emotional problems. Younger female patients experienced more limitations due to physical functioning than older female patients. This finding might indicate that disease-specific uncertainties concerning relationships and offspring are prominent for females in young adulthood, but diminish once a relationship and/or family life are established in later adulthood. Patients with transposition of the great arteries overall seemed to experience a poorer subjective health status, but on the contrary reported the least physical pain. A possible explanation is that past medical experiences put bodily pain in a different perspective. Within the patient sample, displeasure increased, while social inhibition decreased over a 10 year period.

In **chapter 6**, the predictive value of medical variables, covering the complete medical course from birth up till now, for psychopathology (emotional and behavioural problems) in the patient sample at present, were examined. Psychopathology was assessed by the Young Adult Self-Report and Young Adult Behavior Checklist, which were filled in by the patients themselves and other informants respectively. Variables from the adult patient's present life, such as restrictions imposed by physicians, subjective experiences of the scar and low exercise capacity are the best predictors for behavioural and emotional problems as reported by patients themselves. Psychopathology in the patient sample as reported by other informants (e.g. parents, spouses) is best predicted by early hospitalisations (before 1990) with reoperations. Furthermore, being female and the cardiac diagnoses ventricular septal defect and transposition of the great arteries were associated with higher levels of psychopathology. The results show that the level of psychopathology as reported by the patients has a stronger relation with the recent medical status and relevant experiences, whereas the level of psychopathology reported by significant others rather is associated with experiences from the early medical course.

In **chapter 7**, the main findings and conclusions of this thesis were discussed. Overall, the results on psychosocial functioning in the patient sample were satisfactory. Most patients were capable of leading normal lives. However, some difficulties that adult patients with congenital heart disease might encounter were identified. Especially young female patients seem at risk for poor psychosocial functioning. They showed higher levels of psychopathology and poorer subjective experience of physical health. Since these problems seem to diminish when they grow older, these findings indicate that young female patients may face obstacles that are not so prominent in later adulthood. Factors such as subjective experience of the scar, exercise capacity and imposed restrictions, were found to predict behavioural and emotional problems. These issues might raise disease-related uncertainties and worries about entering relationships and having offspring. The results indicate that assessment and treatment of emotional and behavioural problems are desirable in adults with congenital heart disease at an early stage. Several clinical implications following from the results were described.

Samenvatting

Samenvatting

De hoofddoelstelling van dit onderzoek was het onderzoeken van een groot aantal psychosociale aspecten bij volwassenen met een aangeboren hartafwijking. In **hoofdstuk 1**, werd de achtergrond van dit onderzoek geschetst. In de afgelopen decennia hebben, als gevolg van verbeterde medische behandeling, diagnostische en operatietechnieken en daarmee lagere sterftecijfers onder kinderen met een aangeboren hartafwijking, meer patiënten de volwassen leeftijd bereikt. Volwassenen met een aangeboren hartafwijking vormen een nieuwe patiëntengroep. Er is nog maar weinig bekend over hun psychosociaal functioneren. In deze cohort-studie namen 362 volwassenen met een aangeboren hartafwijking deel aan psychologisch onderzoek, 20 tot 33 jaar na hun eerste open hart operatie. Hierbij kwamen verschillende aspecten van psychosociaal functioneren aan bod. Omdat dit onderzoek het tweede vervolgonderzoek van dit cohort betrof, waren we in de gelegenheid om niet alleen naar het huidige functioneren van de patiënten te kijken, maar ook naar de longitudinale ontwikkeling over de afgelopen 10 jaar. De belangrijkste doelstellingen van dit onderzoek waren: (1) het vergelijken van het huidige psychosociaal functioneren van volwassenen met een aangeboren hartafwijking met dat van normgroepen, (2) het bepalen van het longitudinale beloop van psychosociaal functioneren bij volwassenen met een aangeboren hartafwijking, (3) het bepalen van de rol van leeftijd, geslacht en cardiale diagnose ten aanzien van het psychosociaal functioneren van volwassenen met een aangeboren hartafwijking, (4) het bepalen van de voorspellende waarde van een breed aantal medische variabelen op emotionele problemen en gedragsproblemen bij volwassenen met een aangeboren hartafwijking.

In **hoofdstuk 2**, werden biografische kenmerken (bijvoorbeeld zelfstandig wonen, het krijgen van kinderen, opleiding, burgerlijke staat en beroepsniveau), emotioneel functioneren (de schalen Verongelijktheid, Zelfwaardering en Inadequatie van de Nederlandse Persoonlijkheids Vragenlijst) en sociaal functioneren (vrijetijdsactiviteiten) onderzocht. Qua zelfstandig wonen en burgerlijke staat waren patiënten met een aangeboren hartafwijking vergelijkbaar met individuen in de algemene bevolking. Het aantal patiënten met kinderen leek gunstig, maar geen van de 20- tot 24-jarige getrouwde of samenwonende patiënten had kinderen. De onderwijs- en beroepniveaus van de patiënten bleven wat achter bij die in de algemene bevolking, maar deelname aan het arbeidsproces vertoonde een gunstig beeld. De patiënten rapporteerden een goed emotioneel functioneren. Ze gaven blijk van relatief weinig gevoelens van verongelijktheid en neuroticisme en een goed gevoel van eigenwaarde. Vergeleken met de algemene bevolking, lieten de patiënten gunstige vrijetijdsactiviteiten zien. Over het algemeen genomen leken patiënten goed in staat om een normaal leven te leiden en goed gemotiveerd om gebruik te maken van hun mogelijkheden.

In **hoofdstuk 3** werd zowel het huidige niveau van psychopathologie (emotionele problemen en gedragsproblemen) in de patiëntengroep onderzocht, als het longitudinale beloop ervan over een periode van 10 jaar. In het huidige onderzoek vulden de patiënten zelf de Young Adult Self-Report in en een andere informant (bv. ouder of echtgenoot) vulde de Young Adult Behavior Checklist over de patiënt in. Tien jaar geleden werd ook de Young Adult Self-Report afgenomen. Voor patiënten die destijds nog niet de volwassen leeftijd hadden bereikt werden parallelle versies van deze lijst voor de kinderen en adolescenten gebruikt (Child Behavior Checklist in te vullen door ouders en Youth Self-Report in te vullen door adolescenten zelf). Jonge vrouwelijke patiënten vertoonden hogere niveaus van psychopathologie in vergelijking met leeftijdgenoten uit de algemene bevolking. Ouders en partners van de patiënten beoordeelden het gedrag van de patiënten meer afwijkend van de norm dan patiënten zelf. Jongere patiënten vertoonden meer psychopathologie dan oudere patiënten. Ook vertoonden vrouwelijke patiënten meer psychopathologie dan mannelijke patiënten. Verder lieten longitudinale vergelijkingen over een periode van 10 jaar zien dat jongere en vrouwelijke patiënten in de loop van de tijd meer schommelingen vertoonden in het niveau van psychopathologie dan respectievelijk oudere en mannelijke patiënten. De resultaten wijzen op de noodzaak voor beoordeling en behandeling van psychopathologie op jonge leeftijd. Psychopathologie bij jonge vrouwelijke patiënten, en mogelijk onderliggende ziekte-specifieke onzekerheden betreffende het aangaan van relaties en het krijgen van kinderen, verdienen tevens extra aandacht.

In **hoofdstuk 4** werden coping-stijlen (persoonlijke manieren van reageren op problemen) en sociale steun (steun vanuit de sociale omgeving) in de patiëntengroep onderzocht door middel van respectievelijk de Utrechtse Coping Lijst en de Sociale Steun Lijst. Vrouwelijke patiënten vertoonden minder gunstige coping-stijlen in vergelijking met vrouwen uit een normgroep, zoals het minder actief aanpakken van problemen. Mannelijke patiënten vertoonden gunstigere coping-stijlen in vergelijking met mannen uit een normgroep, zoals het vaker zoeken van sociale steun, minder vaak vertonen van passieve reactiepatronen en minder uiten van (negatieve) emoties. In vergelijking met de normgroep, rapporteerde de totale patiëntengroep minder sociale steun te ontvangen, maar ook minder discrepanties te ervaren tussen gewenste en ontvangen sociale steun, wat kan duiden op gevoelens van onafhankelijkheid in deze volwassen patiënten. De resultaten geven aan dat het minder actief aanpakken van problemen bij vrouwen met een aangeboren hartafwijking extra aandacht behoeft.

In **hoofdstuk 5** werd de huidige subjectieve gezondheidstoestand en het longitudinale beloop van psychologisch welbevinden in de patiëntengroep over een periode van 10 jaar onderzocht door middel van respectievelijk de SF-36 en de Medisch Psychologische Vragenlijst voor Hartpatiënten. De patiëntengroep rapporteerde een minder gunstige subjectieve gezondheidstoestand met betrekking tot fysiek functioneren in vergelijking met de normgroep. Daarentegen gaf de patiëntengroep blijk van een beter sociaal functioneren, minder lichamelijke pijn en minder rolbeperkingen als gevolg van emotionele problemen. Jongere vrouwelijke patiënten ervaarden meer beperkingen als gevolg van hun fysiek functioneren dan oudere vrouwelijke patiënten. Deze bevinding zou er op kunnen duiden dat ziekte-specifieke onzekerheden betreffende relaties en het krijgen van kinderen op de voorgrond staan voor vrouwen in de vroege volwassenheid, maar afnemen wanneer een relatie en/of gezinsleven tot stand zijn gekomen in de latere volwassenheid. Patiënten met transpositie van de grote vaten leken over het algemeen een zwakkere subjectieve gezondheidstoestand te ervaren, maar rapporteerden daarentegen het minst vaak lichamelijke pijn. Een mogelijke verklaring hiervoor is dat opgedane medische ervaringen in het verleden lichamelijk pijn in een ander perspectief plaatsen. Binnen de patiëntengroep nam ontstemming toe, terwijl sociale geremdheid afnam over een periode van 10 jaar.

In **hoofdstuk 6** werd de voorspellende waarde van medische variabelen, die het hele medische beloop van de geboorte tot nu bestrijken, voor de huidige mate van psychopathologie (emotionele problemen en gedragsproblemen) in de patiëntengroep onderzocht. Psychopathologie werd gemeten met de Young Adult Self-Report and Young Adult Behavior Checklist, die respectievelijk door de patiënt zelf en een andere informant werden ingevuld. Variabelen afkomstig uit het huidige leven van de volwassen patiënt, zoals beperkingen opgelegd door een arts, het subjectief ervaren van het litteken en inspanningscapaciteit waren de beste predictoren voor psychopathologie zoals gerapporteerd door de patiënten zelf. Psychopathologie in de patiëntengroep zoals gerapporteerd door andere informanten (bijvoorbeeld ouders, echtgenoten) werd het best voorspeld door vroege ziekenhuisopnames (vóór 1990) met reoperaties. Verder werden vrouwen en de cardiale diagnoses ventriculair septum defect en transpositie van de grote vaten geassocieerd met hogere niveaus van psychopathologie. Deze resultaten laten zien dat het niveau van psychopathologie zoals gerapporteerd door de patiënten zelf sterker verband houdt met de recente medische status en relevante ervaringen in vergelijking met het niveau van psychopathologie zoals gerapporteerd door belangrijke anderen, dat meer verband houdt met ervaringen uit het vroege medische verloop.

In **hoofdstuk 7** werden de belangrijkste bevindingen en conclusies van dit proefschrift besproken. Over het algemeen worden bevredigende resultaten gevonden voor psychosociaal functioneren in de patiëntengroep. De meeste patiënten waren in staat een normaal leven te leiden. Echter, er werden enkele moeilijkheden gevonden waar volwassen patiënten met een aangeboren hartafwijking tegenaan zouden kunnen lopen. Vooral jonge vrouwelijke patiënten lopen risico op problemen in het psychosociaal functioneren. Zij vertoonden hogere niveaus van psychopathologie en rapporteerden meer beperkingen als gevolg van hun fysiek functioneren. Daar deze problemen lijken te verminderen wanneer ze ouder worden, kunnen deze resultaten erop duiden dat jonge vrouwelijke patiënten obstakels tegenkomen die in de latere volwassenheid niet meer zo belangrijk zijn. Factoren als het subjectief beleven van het litteken, inspanningscapaciteit en opgelegde beperkingen bleken voorspellers te zijn van emotionele problemen en gedragsproblemen. Deze zaken zouden ziekte-specifieke onzekerheden en zorgen omtrent het aangaan van relaties en het krijgen van kinderen op kunnen roepen. De resultaten wijzen uit dat vroege beoordeling en behandeling van emotionele en gedragsproblemen wenselijk zijn voor volwassenen met een aangeboren hartafwijking. Verschillende uit de resultaten voortkomende klinische implicaties worden beschreven.

Dankwoord
Curriculum Vitae

Dankwoord

Dit onderzoek zou niet mogelijk zijn geweest zonder de medewerking van de vele deelnemers, van wie de meesten voor de tweede maal bereid waren om een dag vrij te maken in hun vaak drukke agenda's en om naar Rotterdam af te reizen. Naast het cardiologisch onderzoeksgedeelte, namen ze ook deel aan het psychologisch onderzoek en gaven antwoord op mijn vele vragen. Ik ben de deelnemers erg dankbaar dat ze, ook buiten de vragenlijsten om, hun ervaringen en verhalen met mij wilden delen. Deze bijzonder waardevolle informatie heeft geleid tot verdieping van dit onderzoek.

De Nederlandse Hartstichting ben ik veel dank verschuldigd voor subsidiëring van dit onderzoek.

Mijn dagelijkse begeleidster en co-promotor dr. Elisabeth Utens, beste Lisbeth, de combinatie van twee Brabantse Elisabeths bleek zeer geslaagd te zijn. Dankjewel dat je altijd tijd voor me had, ook als je thuis druk bezig was met je gezin. Ik heb veel van je geleerd; wanneer mijn aandacht dreigde te verslappen, hield jouw opbouwende kritiek me scherp. Naast het serieuze werk hebben we ook erg veel lol gehad. Bedankt voor je enthousiasmerende begeleiding.

Prof. dr. Frank Verhulst, beste Frank, hartelijk dank dat je mijn promotor wilde zijn. Je commentaar hielp me altijd op de goede weg. Ik heb een erg leuke en leerzame tijd achter de rug op jouw afdeling Kinder- en Jeugdpsychiatrie.

Mijn collega's uit het onderzoeksteam, drs. Jolien Roos-Hesselink, dr. Folkert Meijboom, dr. Ron van Domburg en Willeke van der Bent, bedankt voor de fijne samenwerking en het goede verloop van het onderzoek. Jolien, Folkert en Ron, hartelijk dank voor jullie bijdrage als co-auteurs van mijn artikelen. Ook de medewerkers van de polikliniek Cardiologie, hartelijk dank voor jullie bijdrage aan dit onderzoek.

Alle collega's van de afdeling Kinder- en Jeugdpsychiatrie, met name mijn collega-promovendi en -post-docs Wendy, Gonneke, Ilja, Natasja, Frouke, Mariëlle, Pol, Karen, Jolanda, Alma, Patricia, Kathleen, Nouchka, Victor, Dennis, Mijke en Sabine, hartelijk dank voor jullie gezelligheid, collegialiteit en hulp.

Dr. H.J. Duivenvoorden, hartelijk dank voor uw hulp en suggesties bij hoofdstuk 6. Drs. Jan van der Ende, bedankt voor je hulp bij mijn statistische vragen.

Prof. dr. W.A. Helbing, hartelijk dank dat u secretaris van de leescommissie wilde zijn. Prof. dr. J. Passchier, hartelijk dank voor uw deelname aan de leescommissie. Prof. dr. J.R.T.C. Roelandt, hoofdaanvrager van dit onderzoek, co-auteur van mijn artikelen en lid van de leescommissie, hartelijk dank voor het lezen en beoordelen van mijn proefschrift. Prof. dr. A.J.J.C. Bogers, hartelijk dank voor uw deelname aan de grote commissie en voor het commentaar en de suggesties die u deed als co-auteur. Prof. dr. J. Hess, hartelijk dank voor uw deelname aan de grote commissie.

Mijn familie, vrienden en alle anderen die in de afgelopen jaren steeds vroegen hoe het met het onderzoek en mijn proefschrift ging. Het is af! Jullie warme belangstelling heeft me daar heel erg bij geholpen. Dank jullie wel.

Lieve pap en mam, jullie vertrouwen in mij deed vermoeden dat jullie nooit iets anders dan dit hadden verwacht. Gelukkig heb ik mezelf ook weten te overtuigen. Bedankt voor alle ruimte, belangstelling en steun die ik heb gekregen om volop te studeren en me verder te ontwikkelen. Lieve Paul en Cecile, hoewel we alle drie best verschillend zijn, liggen onze studie- en beroepskeuzes toch dicht bij elkaar. Ik vind het fijn dat we altijd zo betrokken zijn bij elkaar. Dankjewel dat jullie mijn paranimfen willen zijn.

Lieve Dennis, op de eerste dag dat ik aan dit onderzoek begon, werd jij aan mij voorgesteld als de collega met wie ik een kamer zou delen. Gelukkig is het daar niet bij gebleven. Inmiddels delen we een heel fijn leven samen. Het is bijzonder om elkaars promotie van zo dichtbij mee te maken. Heel erg bedankt voor je liefde, vriendschap en steun. Het grootste geluk wat de afgelopen jaren mij hebben gebracht, ben jij!

Curriculum Vitae

Elisabeth Helena Maria van Rijen (roepnaam: Susan) werd op 15 april 1974 geboren te Gilze-Rijen. In 1992 behaalde zij haar V.W.O. diploma aan het Dr. Schaepmancollege te Dongen. In hetzelfde jaar begon zij haar lerarenopleiding aan de Pedagogische Academie Basisonderwijs van de Hogeschool Katholieke Leergangen te Tilburg. Daarnaast begon zij in 1993 in deeltijd aan de opleiding psychologie aan de Katholieke Universiteit Brabant te Tilburg. In 1996 behaalde zij haar onderwijsbevoegdheid voor het basisonderwijs en vervolgde haar studie psychologie in voltijd. In 1999 behaalde zij haar doctoraal psychologie in de richtingen Onderwijs- en Ontwikkelingspsychologie (met specialisatie Kinder- en Jeugdpsychologie) en Psychonomie (met specialisatie Klinische Neuropsychologie). Naast werkzaamheden in het basisonderwijs, was zij tijdelijk werkzaam als psycholoog op Pedologisch Instituut ‘De Hondsberg’ te Oisterwijk.

Vanaf juni 2000 was zij als research-psycholoog aangesteld op de afdeling Kinder- en Jeugdpsychiatrie van het Sophia Kinderziekenhuis – Erasmus MC te Rotterdam (hoofd: Prof. dr. F.C. Verhulst). In deze periode werd een onderzoek uitgevoerd naar psychosociale aspecten van een aangeboren hartafwijking bij volwassen patiënten (projectleider: mw. dr. E.M.W.J. Utens), waarvan de resultaten in dit proefschrift beschreven zijn. Vanaf januari 2003 vervulde zij tevens een post-doc functie in de voorbereiding van een bevolkingsonderzoek naar emotionele problemen en gedragsproblemen.

Sinds oktober 2003 heeft zij een aanstelling als universitair docent ontwikkelingspsychologie aan het Instituut voor Psychologie van de Erasmus Universiteit te Rotterdam (hoofd: Prof. dr. H. Schmidt).