Long-term psychosocial outcomes of children after invasive treatment for congenital heart disease and psychological adjustment of their parents

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Long-term psychosocial outcomes of children after invasive treatment for congenital heart disease and psychological adjustment of their parents

Psychosociale uitkomsten op lange termijn bij kinderen na invasieve behandeling voor een aangeboren hartafwijking en de psychologische aanpassing van hun ouders

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1 Introduction

Introduction

Congenital heart disease

Congenital heart disease (ConHD), the most commonly occurring congenital anomaly, is a structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance (Mitchell et al., 1971).

Historically, surgical treatment of ConHD began in 1939 with procedures outside the heart (Gross & Hubbard, 1939) followed in the 1950s by open-heart surgery with cardiopulmonary bypass. Until about 1970, successful treatment for infants with ConHD was not generally available with a high degree of success.

Over the past decades, advances in diagnostic and surgical techniques and medical treatment of ConHD have gradually evolved and have significantly improved long-term survival of patients with ConHD (Boneva et al., 2001; Eskedal et al., 2005). The number of newborns surviving through infancy, reaching adolescence and adult age has increased enormously (Sparacino, 1994; Wren & O'Sullivan, 2001). While a mortality rate of 85% in the first 20 years of life in the group of patients that require therapy used to be common in the early phases of treatment of ConHD, the 20-years survival has become 85% or better nowadays (Perloff & Warnes, 2001; Perloff, 1991).

The incidence of ConHD is usually estimated by calculating the number of subjects with ConHD per thousand live births. The incidence of ConHD reported in the literature is 6 - 8 per 1000 live births and is in general similar all over the world (Hoffman & Kaplan, 2002). For the Netherlands, it is estimated that every year 1500 infants are born with ConHD. Prevalence refers to the number of affected persons present at any time and represents the difference between the incidence and those who have died. Several studies have attempted to estimate the prevalence of ConHD (Moller et al., 1994; Warnes et al., 2001; Webb et al., 2002; Hoffman et al., 2004). Applying the calculations of Hoffman and colleagues (Hoffman et al., 2004) to the European population of 728 million civilians, the estimation is that between 1.9 and 3.9 million patients with ConHD are currently alive, of which 1.2 to 2.7 million are 15 years or older (Moons et al., 2006). At present, it is estimated that the number of adults with ConHD in the Netherlands could be around 20,000 to 25,000. This number plus an additional 25,000 pediatric patients gives and estimated total of 50,000 patients with ConHD in the Netherlands. Accurate statistics on prevalence rates, however, are lacking.

Despite improved survival, many patients with ConHD may have cardiac residua and sequelae after surgical or interventional treatment. Next to survival, long-term morbidity is of great and improving interest. Morbidity may impair the psychosocial functioning and quality of life of patients with ConHD. Hence, both clinicians and researchers have become more interested in the long-term psychosocial outcomes of patients treated for ConHD.

Psychosocial functioning

In this thesis, the following topics will be discussed. A brief outline of what is known from previous studies is presented here.

Health-related quality of life

Physical problems can hamper the quality of life. Despite the increasing interest in quality of life, consensus is lacking on the definition and measurement of quality of life. Attempts to define quality of life have been guided by the definition of health by the World Health Organization (World Health Organization, 1948). This definition describes health as "a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity". Because health is an important aspect of quality of life, a specific expression has been introduced: health-related quality of life. Health-related quality of life measures have been developed to assess aspects of an individual's subjective experience that relate both directly and indirectly to health, disease, disability, and impairment (Carr et al., 2001) and the effectiveness of treatment.

Assessment of quality of life in children involves specific problems. Children differ from parents in their understanding of health, the causes of illness and its implications (Eiser & Morse, 2001). Children may interpret questions differently and adopt a different time perspective regarding the course of a disease. Their vocabulary and reading skills may be compromised by age and cognitive development. In literature, low concordance has been found between quality of life ratings by children themselves and ratings by proxies, such as parents and health care providers (Sprangers & Aaronson, 1992; Wallander et al., 2001; Theunissen et al., 1998; Verrips et al., 2000; Goldbeck & Melches, 2005). This indicates that both the perspective of the child and of significant others are important in quality of life measurements. Parent-reports can not substitute childreports or vice versa. Until now, little is known about health-related quality of life in children with ConHD. Studies in which the health-related quality of life is rated by both ConHD patients themselves and ratings by their parents are lacking.

Intellectual functioning and school-related behavioural outcomes

In literature, the neurodevelopmental outcome of children with ConHD has received a relatively large amount of attention. Studies have shown that IQ-scores of the majority of children with ConHD were within the normal range (Stavinoha et al., 2003; Forbess et al., 2002a, Forbess et al., 2002b; Wray & Sensky, 2001; Visconti et al., 1999). Neurodevelopmental sequelae, however, are often reported for this population. The neurodevelopmental lags are varied and include motor delays, language and learning disabilities as well as mild deficits in attention (Wypij et al., 2003; Bellinger et al., 2003; Hövels-Gürich et al., 2002b; Forbess et al., 2002b; Bloom et al., 1997; O'Dougherty et al., 1988). The focus of attention

of most of these studies was on neurological abnormalities and intellectual dysfunction, whereas actual long-term schoolfunctioning has been relatively neglected. Studies in which both the long-term intellectual functioning and school-related behavioural outcomes are assessed in a sample of patients with ConHD treated recently, are lacking.

Behavioural and emotional outcomes

Children with (chronic) physical health problems are vulnerable for behavioural and emotional maladjustment (Wallander & Varni, 1998; Lavigne & Faier-Routman, 1992). Research has shown that children and adolescents with ConHD exhibit significantly more behavioural and emotional problems than children from the general population (Fredriksen et al., 2004; Hövels-Gürich et al., 2002a; Janus & Goldberg, 1995; Utens et al., 1993), irrespective of the severity of the disease (Fredriksen et al., 2004; Utens et al., 1993). Mainly internalising problems were reported, such as feelings of anxiety (Gupta et al., 1998 and 2001; Utens et al., 1993; Kramer et al., 1989), depression (Gupta et al., 1998 and 2001; Utens et al., 1993), and inferiority (Kramer et al., 1989). To a lesser extent, externalising problems in children and adolescents with ConHD were reported (Kramer et al., 1989).

Medical predictors

It is important to know to what extent factors associated with the medical course in early childhood are predictive for long-term behavioural and emotional problems. Aspects in the treatment and support of children with ConHD that need special attention can then be identified. Previously, several predictors for behavioural and emotional problems in children with ConHD have been identified, varying from maternal perceptions (DeMaso et al., 1991) to medical variables such as age at surgical repair, deep hypothermic circulatory arrest, and number of heart operations (Utens et al., 1998; Bellinger et al., 1997). Studies in which medical predictors of long-term behavioural and emotional problems are assessed in a sample of children treated for ConHD recently, however, are lacking.

Psychological distress and coping styles of parents

The diagnosis of ConHD has important implications for parents and the family as a whole. In an empirical study, Kirschenbaum Cohn (1996) investigated reactions of parents to the diagnosis that their infant had a ConHD. They found that these parents showed more fear, anger and sadness compared to parents of healthy infants. Parents of children with ConHD have been reported to express more distress and hopelessness than parents of healthy children or parents of children with other chronic conditions (Lawoko & Soares, 2002; Uzark & Jones, 2003a; Gardner et al., 1996; Goldberg et al., 1990a). Furthermore, Davis et al. (1998) found that maternal adjustment in mothers with ConHD children was influenced by high levels of daily stress and by coping techniques and was not significantly associated with the severity of the cardiac defect.

Previously, several studies conducted on parents of children with ConHD focused on parental adjustment to the diagnosis and the short-term impact of the illness on the family, the early mother-infant relationship, and care-giving problems (Svavarsdottir & McCubbin, 1996; Goldberg et al., 1990a; Goldberg et al., 1991; Lobo, 1992). Reactions of parents to an illness identified in infancy may be quite different from those occurring when children are older. Until now little is known about coping styles in parents of children with ConHD. The few studies executed, assessed styles of coping in parents prior to or shortly after cardiac surgery (Utens et al., 2000a, 2002; Wray & Sensky, 2004). Long-term coping styles may differ from those shortly after the disease was discovered. Nowadays, the long-term survival of children with ConHD has improved, but parents still have to cope with the effects of the illness on the functioning, development, and quality of life of their child, irrespective of the severity of the disease. Data on the long-term effects of ConHD on parents, however, are lacking.

The present study: a long-term follow-up

The aim of the study described in this thesis is to investigate the long-term psychosocial outcomes of children and adolescents who underwent invasive treatment for ConHD between 1990 and 1995 in the Erasmus University Medical Centre Rotterdam. Children and adolescents of four cardiac diagnostic groups were examined with standardized instruments. Furthermore, the psychological adjustment of parents of the present patient sample was studied. Assessment of psychosocial functioning may identify important issues in the child or their parents that are not readily apparent in the routine clinical evaluations.

Various indicators of psychosocial functioning of children and adolescents with ConHD were measured, including health-related quality of life, intellectual and school-related behavioural functioning, parent- and self-reported behavioural and emotional functioning. Coping styles and psychological distress in parents of patients were also investigated. Outcomes on psychosocial functioning in the patient sample were compared with those in the normal population. The role of gender, age, and cardiac diagnosis was examined systematically. Furthermore, the predictive value of a wide range of medical variables on long-term behavioural and emotional problems in the patient sample was examined. Finally, a historical comparison was made to compare the level of long-term behavioural and emotional problems in the present patient sample, treated recently, versus that of a comparable, historical sample of same-aged patients operated for ConHD before 1980 in the Erasmus University Medical Centre Rotterdam.
The aims of the present study were:

1. To compare the present psychosocial functioning of children and adolescents who underwent invasive treatment for ConHD at a young age with that of normative samples.
2. To determine the role of gender, age, and cardiac diagnosis on the psychosocial functioning of children and adolescents with ConHD.
3. To compare the level of long-term behavioural and emotional problems of the present sample of children and adolescents with ConHD treated recently, that is between 1990 and 1995, with that of a historical sample of children and adolescents operated for ConHD between 1968 and 1980 in the Erasmus University Medical Centre Rotterdam.
4. To determine the predictive value of a wide range of medical variables on long-term behavioural and emotional problems in children and adolescents with ConHD.
5. To compare the levels of psychological distress and current coping styles of both mothers and fathers of children and adolescents from the patient sample with that of reference groups.

Methods

Inclusion criteria

During the follow-up, which took place in 2003-2004, consecutive surviving patients of 4 diagnostic groups, who underwent their first invasive treatment for ConHD at least 7 years and 6 months ago (between 1 January 1990 and 1 January 1996) in the Erasmus University Medical Centre Rotterdam, and who were younger than 15 years at the time of the treatment, were eligible. The sample in this study encompassed the following cardiac diagnostic groups: atrial septal defect (ASD), ventricular septal defect (VSD), transposition of the great arteries (TGA) and pulmonary stenosis (PS). Patients with proven syndromes of mental retardation including Down syndrome were excluded.

Patient sample

The target population consisted of 246 consecutive surviving patients. At follow-up 40 patients were lost (23 moved abroad, 17 were untraceable). The remaining 206 patients were aged 7-28 years. Of this group 35 patients were aged 18-28 years. Results of the participating 18-28-year-old patients were not included in this thesis. The present patient sample consisted of 171 patients, aged 7-17 years, and their parents. Since the instruments used to assess various indicators of psychosocial functioning did not cover the same age-ranges, the number of patients and parents in the chapters 2 to 7 of this thesis differ. The exact numbers of patients and parents are indicated in the corresponding chapters.

Instruments

In selecting assessment instruments we choose to use (where possible) internationally well-known standardized assessment instruments of which the psychometric properties (reliability, validity) have been proven to be satisfactory. During the psychological examination, the TNO-AZL Child Quality of Life Questionnaire (TACQOL; Vogels et al., 2000) was used to assess health-related quality of life. Intellectual functioning was assessed with the Dutch version of the Wechsler Intelligence Scale for Children-Revised (WISC-R; Van der Steene et al., 1986). Behavioural and emotional problems were assessed with the Child Behavior Checklist (CBCL; Achenbach, 1991a), the Youth Self-Report (YSR; Achenbach, 1991b) and the Teacher's Report Form (TRF; Achenbach, 1991c). Psychological distress of parents was assessed by the General Health Questionnaire (GHQ; Koeter & Ormel, 1992). The Utrecht Coping List (UCL; Schreurs et al., 1993) was used to assess coping styles of parents. A detailed description of these instruments can be found in the chapters 2 to 7 of this thesis. Furthermore, a semi-structured interview with parents was used to assess biographical variables such as living conditions, offspring, education, occupational status (Rotterdams Kwaliteit van Leven - Interview; Utens et al., 2000b).

Assessment procedure

The research protocol was approved by the Dutch Central Committee on Medical Research involving Human Subjects before the start of the study. All patients were traced and approached uniformly. After an information letter was received, patients were called for an appointment by the research assistant. Before participating in the study, parents and/or patients signed an informed consent and returned it by mail. The definite cardiac diagnosis was checked by a paediatric cardiologist (WH). During the visit to the Erasmus University Medical Centre Rotterdam, patients were interviewed and tested by a psychologist (AS) and medically examined by a physician/cardiologist. Parents of patients completed the questionnaires independently from their child in the waiting room. Parents were interviewed after the psychological examination of their child.

The structure of this thesis

The aim of this thesis is to provide insight into long-term psychosocial functioning of a sample of children and adolescents with ConHD treated recently, that is between 1990 and 1995. In addition, the psychological adjustment of their parents was assessed.

In *chapter 2*, health-related quality of life is investigated in children and adolescents from the present patient sample and their parents. In *chapter 3*, the level of intellectual functioning of the patients was examined. School-related behavioural and emotional outcomes of the patients, reported by their teachers, are described. In *chapter 4*, the

occurrence of a wide range of behavioural and emotional problems in the patient sample is assessed. In *chapter 5*, a historical comparison is made between the long-term behavioural and emotional outcomes of the present patient sample versus a comparable, historical sample of children and adolescents treated for ConHD between 1968 and 1980. In *chapter 6*, medical predictors covering the medical course from birth up till now, for long-term behavioural and emotional problems in the present patient sample, are identified. In *chapter 7*, the level of psychological distress and coping styles in both mothers and fathers from the present patient sample were examined. Finally, in *chapter 8*, the main findings and conclusions of this thesis are discussed. Implications and recommendations for medical practice are given.

2 Health-related quality of life in children and adolescents after invasive treatment for congenital heart disease

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Abstract

Background

Since the 1980s treatment techniques for congenital heart disease (ConHD) have gradually evolved. Therefore, actual information on the outcomes, including quality of life is required. Health-related quality of life was assessed long-term in 4 diagnostic groups of children, who underwent invasive treatment for ConHD between 1990 and 1995.

Methods

The scores on the TNO-AZL Child Quality of Life Questionnaire (TACQOL) of both children with ConHD and their parents were compared with those of a same-aged reference group.

Results

The total sample of ConHD children (N=113, 8-15 years old) obtained significantly lower mean scores on motor functioning, cognitive functioning, and positive emotional functioning than reference peers, reflecting an experience of poorer functioning.
ConHD children, aged 8-11 years, obtained lower mean scores on 5 of the 7 TACQOL scales than reference peers. They also had a lower score on positive emotional functioning than 12-15-year-old ConHD children.
The total sample of ConHD children obtained lower outcomes compared to their parents on 4 of the 7 TACQOL scales.
No significant differences were found in health-related quality of life between ConHD boys and girls, neither between different diagnostic groups.

Conclusion

Overall, this sample of recently treated ConHD children showed a worse health-related quality of life compared to reference groups. These findings deserve further attention.

Introduction

Mortality in children with congenital heart disease (ConHD) has substantially decreased over the last four decades, as a result of important advances in diagnostic, surgical and catheter interventional techniques (Boneva et al., 2001). The age at surgical intervention has been reduced significantly for most conditions. New techniques, including catheter intervention, have been introduced for several congenital heart conditions. Furthermore, improved techniques have allowed repair of congenital heart anomalies previously considered inoperable.

Traditionally, mortality and morbidity have been viewed as the key outcomes when considering the efficacy of medical interventions. It has become increasingly clear that improved long-term survival may result in long-term morbidity (Gatzoulis et al., 1999, 2000; Meijboom et al., 1994; Meijboom et al., 1996; Mair et al., 2001). This may impair quality of life. Accordingly, quality of life recently has become an important outcome measure, in addition to morbidity and mortality.

Despite the increasing interest in quality of life, both in research and clinical practice, consensus is lacking on the definition and measurement of quality of life. The World Health Organization defined health as "a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity" (World Health Organization, 1948). This definition has been highly influential in defining the concept of quality of life.

Sometimes the terms health status and health-related quality of life (HRQol) seem to be used as equivalents (Vogels et al., 2000). Health status refers to assessment by a person of his or her actual, more objective problems and limitations in functioning, whereas HRQol includes the person's subjective, emotional evaluation and reaction to such problems and limitations. In this study HRQol is considered as a multidimensional construct that encompasses the dimensions described by the World Health Organization.

Until now, little is known about HRQol in children with ConHD (Uzark et al., 2003). The few studies (Culbert et al., 2003; Dunbar-Masterson et al., 2001; Kendall et al., 2001) executed in this field often showed methodological problems, such as use of limited samples (e.g. only one cardiac diagnostic group), and variation in methods across studies. In general, it is known that children and parents may have different views about the impact of illness. Studies in which the HRQol is assessed by both children and parent ratings in a large sample of children with ConHD treated recently are lacking.

The present study is part of a follow-up study concerning the long-term medical and psychological outcomes in children, adolescents and young adults who underwent invasive treatment for ConHD recently, that is between 1990 and 1995.

The aims of the present study were:

1. To compare the level of HRQoI in 8-15-year-old children and adolescents who underwent invasive treatment for ConHD at young age with that of reference groups.
2. To identify the role of sex, age and cardiac diagnosis on the level of HRQoI for this sample.
3. To determine the agreement between child and parent ratings within this sample.

Methods

TNO-AZL Child Quality of Life Questionnaire

The TNO-AZL Child Quality of Life Questionnaire (TACQOL) is a generic instrument, designed to assess general aspects of HRQoL of children aged 8 to 15 years (Vogels et al., 2000). It assesses the occurrence of functional problems, and if such a problem occurs, negative emotional reactions are assessed, too. Two parallel questionnaires are available with parallel items: a child form and a parent form. The child form is identical to the parent form except that the child form is worded in the first person.

In the TACQOL the construct of HRQoI is operationalised by 7 dimensions. Each dimension is assessed by a scale consisting of 8 items: pain and physical symptoms, motor functioning, autonomy, cognitive functioning, social functioning, global positive emotional functioning and global negative emotional functioning (see Figure 1).

Figure 1. Items of the TACQOL parent form

Pain and physical symptoms	
Has your child had	Earaches or sore throats; Stomach aches or abdominal pain; Headaches; Dizzy; Felt sick/nauseous;
Was your child	Tired; Sleepy; Dozy/lethargic
Motor functioning	
Did your child have difficulty with	Running; Walking; Standing; Walking downstairs; Playing; Running or walking long distances; Balance; Doing things handily or quickly
Autonomy	
Did your child have difficulty with	Going to school on his own; Washing himself; Getting dressed on his own; Eating or drinking on his own; Sports or going out to play on his own; Doing hobbies on his own; Riding a bicycle
Cognitive functioning	
Did your child have difficulty with	Paying attention, concentrating; Understanding schoolwork; Understanding what others said; Arithmetic; Reading; Writing; Learning; Saying what he meant
Social functioning	
My child was	Able to play or talk happily with other children; Able to stand up for himself with other children; Other children asked to play with them; At ease with other children; Able to play or talk happily with parents; Incommunicative or quiet with parents; Restless or impatient with parents; Defiant with parents
Positive emotional functioning	
My child felt	Joyful; Relaxed; In good spirits; Happy; Contented; Cheerful; Enthusiastic; Confident
Negative emotional functioning	
My child felt	Sad; Aggressive; Angry; Shorttempered; Worried; Jealous; Gloomy; Anxious

HRQol is defined as health status weighted by the subjective, emotional impact of problems in health status. Therefore, in all scales (except for the two scales concerning emotional functioning) each item consists of two questions. The TACQOL first assesses the frequency of occurrence of a specific complaint or limitation. If such a problem exists, then the subjective appraisal of this problem is assessed. The reference period is formulated as 'the last few weeks'.

Figure 2 shows an example of such a question. Each item is encoded into a single score ranging from 0 to 4 (see Figure 2). The range of the sum scores varies between 0 to 32. The item scores on emotional problems were on a 0 to 2 scale and the scale scores ranged from 0 to 16. For all TACQOL scales a higher score indicates a better HRQol. As HRQol is defined as a multidimensional construct, no total score is calculated. The satisfactory psychometric properties (reliability, validity) of the TACQOL child and parent form have been described in detail by Verrips et al. (1999).

Figure 2. Item-example of the TACQOL parent form (scoring in parentheses)

Pain and physical symptoms

Try to remember how your child was in recent weeks …

Has your child had earaches or sore throats?

❑ never *(4)* ❑ occasionally ❑ often

At that time, my child felt:

❑ fine *(3)* ❑ not so good *(2)* ❑ quite bad *(1)* ❑ bad *(0)*

Inclusion and exclusion criteria

During the follow-up, which took place in 2003-2004, consecutive surviving patients of 4 diagnostic groups, who underwent their first invasive treatment for ConHD between 1 January 1990 and 1 January 1996 in the Erasmus University Medical Centre Rotterdam, and who were younger than 15 years at the time of the treatment, were eligible. The sample in this study encompassed the following cardiac diagnostic groups: atrial septal defect (ASD), ventricular septal defect (VSD), transposition of the great arteries (TGA) and pulmonary stenosis (PS). Patients with proven syndromes of mental retardation including Down syndrome were excluded.

Patient sample

The target population consisted of 246 consecutive surviving patients. At follow-up 40 patients were lost (23 moved abroad, 17 were untraceable). For the present study, 154 patients aged 8 to 15 years (upper age-range of the questionnaire) and their parents were eligible. 41 Children and 40 of their parents refused to participate (26 on practical and 14 on emotional grounds). The total group non-responders for the patient sample consisted of 56% boys (n=23) and 44% girls (n=18) with a mean age of 11 years. The final study sample consisted of the following subsamples of patients between 8-15 years and their parents:

1. TACQOL patient sample.
   A total of 61 boys and 52 girls (n=113) provided usable TACQOL child forms. The response rate of this sub sample, corrected for persons lost to follow-up, was 73%.
2. TACQOL parent sample.
   Parents of 61 male and 53 female patients (n=114, 78% mothers and 22% fathers) provided usable TACQOL parent forms. The corrected response rate of this sub sample was 74%.

Numbers of patients belonging to each cardiac diagnostic group of the patient sample were: surgical closure of ASD: n=22 (mean age 13 years, SD=1.6); surgical closure of VSD: n=46 (mean age 11 years, SD=1.8); arterial switch operation of TGA: n=33 (mean age 10 years, SD=1.5); and balloon dilatation for PS: n=12 (mean age 12 years, SD=2.4).

Reference groups

Data of the patient sample were compared with data derived from Dutch normative samples from the general population, with comparable age-ranges published in the manual (Vogels et al., 2000). Twelve regional Centres for Preventive Youth Care spread over the Netherlands selected from their registries a random sample of children from the general population, stratified by gender and age. Normative data were available for two categories: 8-11 and 12-15 years.

1. TACQOL child form reference group.
   For ages 8-11 years, the reference group consisted of 1078 children (534 boys and 544 girls).
   For ages 12-15 years the number of children differs for almost all TACQOL scales (for exact numbers, see table 2).
2. TACQOL parent form reference group.
   Normative data for the TACQOL parent form are only available for the age-range 8-11 years. The reference group consisted of 1094 parents (of 543 male and 551 female children).

Assessment procedure

The research protocol was approved by the ethics committee review board before the start of the study. All patients were traced and approached uniformly. After an information letter was sent patients were called for an appointment by the research assistant. Parents and/or patients signed an informed consent and returned it by mail before participating in the study. The definite cardiac diagnosis was checked by a paediatric cardiologist (W.H.). During the visit to the Erasmus University Medical Centre, patients were interviewed and tested by a psychologist (A.S.) and medically examined by a cardiologist. The TACQOL child form was filled in by 8-15-year-old patients during the psychological examination. Parents of patients completed the TACQOL parent form independently from their child in the waiting room. Three patients and six parents preferred to complete the TACQOL child form respectively the parent form at home and returned the questionnaires in a prepaid envelope.

Statistical analyses

All analyses were performed with two-sided tests; $p<0.05$ was considered significant. One sample t-tests were used to test differences in group means between the patient sample and reference group on the TACQOL scales. For these analyses, Cohen's D's (Cohen, 1988) were computed to assess the magnitude of differences in mean scores between the patient sample and the reference group. According to Cohen's (Cohen, 1988) criteria, a standardised difference of 0.20 can be considered as small, 0.50 as medium, and 0.80 as high.

To identify the role of sex (boys versus girls), age (8-11 versus 12-15 years) and cardiac diagnosis (ASD, VSD, TGA, PS) within the patient sample, univariate analyses of variance (ANOVAs) were performed on all TACQOL scales. According to Cohen (Cohen, 1988), effects accounting for 1.0-5.9% of variance are considered small, 5.9-13.8% medium and >13.8% large.

To investigate agreement between different informants (parents and children) respectively to test differences between informants, Pearson correlations and MANOVAs with repeated measures were performed on the TACQOL scales of 8-15-year-old patients with both a parent and child form completed.

Results

Comparison of the total ConHD sample versus reference groups

Table 1 shows the mean scores on the seven TACQOL scales, by each sex, for the ConHD patient and the reference group, as reported by children aged 8-15 years and their parents. It also shows significant differences and the corresponding effect sizes (Cohen's D).

Table 1. Mean scores, standard deviations and Cohen's D on the TACQOL child and parent form for the ConHD sample and reference group, for ages 8-15 years

	TACQOL Child form												
	Boys				Girls				All				
	ConHD		Reference		ConHD		Reference		ConHD		Reference		
Scales	**M (n=61)**	**SD**	**M (n)**	**SD**	**M (n=52)**	**SD**	**M (n)**	**SD**	**M (n=113)**	**SD**	**M (n)**	**SD**	**Cohen's D**
Pain and physical symptoms	24.7	4.8	24.9 (1125)	5.0	23.7	5.1	23.6 (1202)	5.5	24.2	5.0	24.3 (2327)	5.3	0.01
Motor functioning	28.3*	4.2	30.0 (1127)	3.2	27.3*	5.6	29.6 (1203)	3.3	27.8*	4.9	29.8 (2330)	3.2	0.6
Autonomy	30.3	2.6	NA	NA	30.2	3.3	NA	NA	30.3	2.9	NA	NA	NA
Cognitive functioning	26.8*	4.5	28.1 (1127)	3.9	26.4*	4.4	27.9 (1203)	4.2	26.6*	4.4	28.0 (2330)	4.1	0.3
Social functioning	28.6	3.1	NA	NA	28.2	4.1	NA	NA	28.4	3.6	NA	NA	NA
Positive emotional functioning	12.9	2.8	13.3 (1122)	2.6	12.3*	3.0	13.3 (1202)	2.7	12.6*	2.9	13.3 (2324)	2.7	0.2
Negative emotional functioning	12.0	3.5	11.7 (1122)	2.7	11.3	2.6	11.6 (1202)	2.6	11.7	3.1	11.6 (2324)	2.6	0.02

	TACQOL Parent form					
	Boys		Girls		All	
	ConHD		ConHD		ConHD	
Scales	**M (n=61)**	**SD**	**M (n=53)**	**SD**	**M (n=114)**	**SD**
Pain and physical symptoms	27.1	4.2	25.4	5.8	26.3	5.1
Motor functioning	29.9	3.3	29.4	4.7	29.7	4.0
Autonomy	31.2	2.4	31.0	2.4	31.1	2.4
Cognitive functioning	26.4	6.0	27.9	3.9	27.1	5.2
Social functioning	28.8	4.2	29.0	3.5	28.9	3.9
Positive emotional functioning	13.5	2.8	13.7	2.9	13.6	2.8
Negative emotional functioning	11.7	2.8	11.5	2.2	11.6	2.5

* Significant ($p < 0.05$) difference.
NA=Not available

Compared with the normative group, the total sample of ConHD children obtained significantly lower scores on motor functioning ($p<0.001$), cognitive functioning ($p=0.001$), and positive emotional functioning ($p=0.016$), indicating poorer functioning. Motor functioning, and cognitive functioning appeared to be significantly lower in ConHD boys ($p=0.002$ respectively $p=0.021$) and ConHD girls ($p=0.004$ respectively $p=0.024$) compared to same-sex peers from the general population. ConHD girls also reported a significantly lower score on positive emotional functioning ($p=0.025$) than reference girls did.

Comparison of two age groups of ConHD children and their parents with reference groups

Table 2 shows the mean scores on all TACQOL scales for two age categories (8-11 and 12-15 years) for the patient and the reference group as reported by children and parents. Compared with same-aged peers from the general population, ConHD children, aged 8-11 years, obtained significantly lower mean scores on 5 of the 7 TACQOL scales (all $p<0.005$): motor functioning, autonomy, cognitive functioning, social functioning, and positive emotional functioning. No differences were found on the scales pain and physical symptoms and negative emotional functioning. For ConHD children, aged 12-15 years, however, only one significant difference was found. Motor functioning ($p=0.024$) appeared to be significantly lower compared to same-aged reference peers.

Parents of ConHD children aged 8-11 years reported significantly lower scores for their children on cognitive functioning ($p=0.002$) and on positive emotional functioning ($p=0.006$) than parents from the general population for their children.

Comparisons within the patient sample: effects of sex, age, and cardiac diagnosis

ConHD boys did not differ from ConHD girls on any of the TACQOL child form scales. Neither any difference was found on parent-reports between parents of ConHD boys versus girls.

ConHD children, aged 8-11 years, had a significantly lower mean score on positive emotional functioning ($p=0.003$) than ConHD children aged 12-15 years. Parents of ConHD children aged 8-11 years reported significantly lower scores on cognitive functioning ($p=0.037$) than parents of ConHD children aged 12-15 years.

On the parent form, a significant interaction effect of sex and age group was found on cognitive functioning ($p=0.005$). This medium effect (according to Cohen's criteria (Cohen, 1988)) indicated that the difference between parent-reports on their 8-11-year-old ConHD boys versus girls was larger than that of parent-reports regarding their 12-15-year-old ConHD boys versus girls.

Table 3 shows the mean scores on the seven TACQOL scales for different cardiac diagnostic groups and their parents. No significant differences for cardiac diagnosis were

Table 2. Mean scores and standard deviations of two age groups on the TACQOL child and parent form for the ConHD sample and reference group

	Ages 8-11				Ages 12-15				Ages 8-11				Ages 12-15	
	Child form				Child form				Parent form				Parent form	
	ConHD n=58		Reference n=1078		ConHD n=55		Reference		ConHD n=58		Reference n=1094		ConHD n=56	
Scales	**M**	**SD**	**M**	**SD**	**M**	**SD**	**M (n)**	**SD**	**M**	**SD**	**M**	**SD**	**M**	**SD**
Pain and physical symptoms	24.4	4.7	24.9	5.1	24.0	5.3	23.7 (1249)	5.4	27.0	4.1	26.9	4.0	25.6	5.9
Motor functioning	27.6[1]	4.3	29.8	3.2	28.1[1]	5.5	29.8 (1252)	3.3	29.7	3.6	30.6	2.7	29.6	4.4
Autonomy	30.1[1]	2.9	31.2	1.9	30.6	2.9	NA	NA	31.1	1.8	31.3	1.6	31.1	2.9
Cognitive functioning	26.1[1]	4.5	28.5	3.9	27.1	4.3	27.6 (1252)	4.1	26.1[1,2]	5.9	28.7	3.9	28.1[2]	4.0
Social functioning	27.8[1]	3.9	29.7	2.8	29.0	3.1	NA	NA	29.0	3.1	29.7	2.7	28.8	4.5
Positive emotional functioning	11.8[1,2]	3.0	13.6	2.5	13.4[2]	2.5	13.0 (1246)	2.8	13.7[1]	2.7	14.7	2.2	13.5	3.0
Negative emotional functioning	11.4	3.7	11.6	2.7	12.0	2.5	11.6 (1246)	2.6	11.6	2.4	11.5	2.5	11.7	2.7

[1] Significant ($p < 0.05$) difference between ConHD sample and corresponding reference group.

[2] Significant ($p < 0.05$) difference between scores on the child or parent form of younger (8-11 years) versus older (12-15 years) ConHD patients.

NA=Not available

Table 3. Mean scores and standard deviations on the TACQOL child and parent form for different cardiac diagnostic groups

	TACQOL Child form									TACQOL Parent form								
	ASD n=22		VSD n=46		TGA n=33		PS n=12			ASD n=22		VSD n=48		TGA n=32		PS n=12		
Scales	**M**	**SD**	**M**	**SD**	**M**	**SD**	**M**	**SD**	**Main Effects**	**M**	**SD**	**M**	**SD**	**M**	**SD**	**M**	**SD**	**Main Effects**
Pain and physical symptoms	23.7	5.7	24.6	4.6	24.2	5.3	23.8	4.8	ns	24.2	7.0	26.5	4.9	27.4	3.8	26.4	3.8	ns
Motor functioning	27.6	6.4	27.6	5.0	27.7	4.3	29.2	2.8	ns	29.6	5.9	29.6	3.6	29.8	3.3	30.0	3.1	ns
Autonomy	30.3	3.8	30.3	2.4	30.0	3.4	31.0	1.4	ns	31.1	3.0	30.9	2.7	31.4	1.3	30.8	2.3	ns
Cognitive functioning	26.2	5.3	26.3	3.9	27.3	4.6	26.5	4.2	ns	29.1	3.9	27.1	4.2	26.0	6.5	26.7	6.2	ns
Social functioning	28.6	3.1	27.9	4.2	28.7	3.4	29.4	2.1	ns	29.3	3.9	28.5	4.7	29.4	2.3	28.4	3.5	ns
Positive emotional functioning	13.5	2.8	12.5	2.8	12.0	3.2	13.2	2.1	ns	14.1	2.5	13.7	2.9	13.1	2.8	13.7	3.1	ns
Negative emotional functioning	12.2	2.1	11.3	2.7	11.7	4.4	12.0	2.4	ns	11.6	2.5	11.4	2.7	11.6	2.4	12.5	2.1	ns

Note. Abbreviations used are: ASD = Atrial Septal Defect, VSD = Ventricular Septal Defect, TGA = Transposition of the Great Arteries, PS = Pulmonary Stenosis

found. Neither any difference was found between mean scores on TACQOL child and parent form scales of children treated by surgical intervention (ASD, VSD and TGA) and treated by catheter intervention (PS).

Comparison between different informants: ConHD children versus their parents

Pearson correlation were calculated between all TACQOL scales of the parent and child form. All correlations were significant at a significance level of $p<0.01$. 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 0.50 are considered large. The Pearson correlation coefficients ranged from 0.26 to 0.51. For motor functioning a large correlation was found between child and parent-reports. Medium correlations were found between pain and physical symptoms, social functioning and positive emotional functioning as reported by children and their parents.

The total sample of 8-15-year-old ConHD children reported significantly poorer quality of life than their parents, on four of the seven scales: pain and physical symptoms ($p<0.001$), motor functioning ($p<0.001$), autonomy ($p=0.006$) and positive emotional functioning ($p=0.001$).

A significant interaction effect of informant and age was found for positive emotional functioning indicating that the difference between child and parent-reports of children aged 8-11 years is larger than that between older children and their parents.

Discussion

HRQol in the patient sample compared to the reference group

The results of the present study showed significantly lower HRQol scores regarding motor functioning, cognitive functioning, and positive emotional functioning for the total sample of ConHD children compared to the reference group, reflecting an experience of poorer functioning in these domains for ConHD children. ConHD children, aged 8-11 years, reported significantly poorer quality of life than reference peers on 5 domains: motor functioning, cognitive functioning, autonomy, social functioning and positive emotional functioning. Their parents reported significantly poorer quality of life regarding their children for cognitive functioning and positive emotional functioning compared to parents from the reference group. In contrast, reports of 12-15-year-old ConHD patients were comparable to those of reference peers (except on motor functioning), which is a favourable outcome. Overall, from our results it can be concluded that especially 8-11-year-old ConHD children are at risk for poorer HRQol, both according to their parents' as to their self-reports.

Our findings confirm those of Uzark et al. (2003), who found that ConHD children functioned less well than healthy controls regarding psychological health, emotional, social and school functioning, both according to self-reports and parent-reports. Our study methods differed from theirs: their study sample consisted of children aged 5-18 years (n=250) and parents of children aged 2-18 years (n=344). In contrast with our findings, Culbert et al. (2003) reported a better self-perceived health status on the Child Health Questionnaire for 11-15-year-old children with TGA, in comparison to the normative population. The instrument used in their study, however, was more focused on health status and covered different dimensions. Their sample consisted of child-respondents alone, who had undergone an arterial switch, atrial switch or Rastelli operation between 1985 and 1989, which makes it difficult to compare findings.

Our study showed that poorer HRQol regarding cognitive and motor functioning was reported by the total sample of ConHD children, and regarding cognitive functioning by parents of 8-11-year-old patients. Our unfavourable finding regarding motor functioning is in line with literature (Sticker, 2004). Previous studies showed unfavourable neurodevelopmental outcomes for certain cardiac defects (Bellinger et al., 1999, 2003; Wernovsky et al., 2000; Mahle, 2001; Mahle et al., 2000). Bellinger et al. (1999, 2003), for example, reported that neurocognitive performance in children with TGA, operated with circulatory arrest and low flow, was below expected in several domains, including IQ, expressive language, visual-motor integration, motor planning and organization. Results between studies are difficult to compare since samples and instruments used, differ.

The items of the TACQOL scale cognitive functioning assess the emotional reactions of having difficulties with concentrating, understanding schoolwork, understanding what others say, arithmetic, reading, writing, learning, and in saying what you mean (see Figure 1). From our study one might suggest that neurocognitive sequelae in ConHD children may have implications for a poorer HRQol in this domain. However, our results are remarkable, considering other studies in which no neurodevelopmental delay was found for diagnosis such as ASD and VSD. Visconti et al. (1999) compared in their study a group of children who underwent closure of an ASD by surgery (n=26) with another group of children who underwent ASD closure by a catheter-delivered device (n=19). They found that children's IQ and achievement scores were in the normal range for both groups. In the study of Oates et al. (1995a), no significant difference in IQ scores was found between a group of children operated for tetralogy of Fallot, TGA, VSD and ASD and that IQ scores were in the normal range.

In a follow-up study, Dunbar-Masterson et al. (2001) found that parents reported more problems with attention, learning, and speech, as well as greater frequency of developmental delay for their children with TGA. Overall, the physical and psychosocial health status of patients as measured by the Child Health Questionnaire was similar to that in

the general population. The study methods of Dunbar-Masterson et al. (2001), however, differed from ours, since they used only one diagnostic group (TGA) and measured general health status of ConHD children aged 8 years, as reported by parents only, whereas our instrument assessed HRQol as reported by both ConHD children themselves and their parents. A strength of our study is that the emotional impact of difficulties on different quality of life domains was assessed by direct reports from children themselves and their parents, whereas most of previous studies (Culbert et al., 2003; Dunbar-Masterson et al., 2001) focused on health status and used one source of information (parents of children or children themselves).

Sex, age, and cardiac diagnosis within the patient sample

In this study HRQol of boys and girls with different cardiac diagnosis is similar. Culbert et al. (2003) found in their study that male gender was associated with unfavourable outcomes in emotional and behavioural areas, and female gender was associated with lower general health perceptions. In the general population Theunissen et al. (1998) found no clear effect of gender on quality of life outcomes for children.

As to age we found in this study that ConHD children, aged 8-11 years, showed a lower HRQol on positive emotional functioning than children aged 12-15 years. As far as we know, no previous study has reported about this. Parents of ConHD children aged 8-11 years reported poorer quality of life regarding cognitive functioning than parents of ConHD children aged 12-15 years.

In this study, no significant differences were found as to the HRQol of the different cardiac diagnostic groups. This is in line with previous findings, in which neither differences were found between the level of emotional adjustment (psychopathology) of different cardiac diagnostic groups (Utens et al., 1993). Next to cardiac diagnosis, several other variables such as length of time since first invasive treatment, could have influenced the HRQol of this patient sample. Further analyses (linear regression) showed that length of time since first invasive treatment was not significantly related to HRQol at follow-up. Future research should investigate which medical variables are significant predictors of long-term HRQol at follow-up.

Agreement between child and parent rating

The total sample of ConHD children reported significantly poorer quality of life than their parents did about them regarding pain and physical symptoms, motor functioning, autonomy and positive emotional functioning. The difference between child and parent-reports of children aged 8-11 years was larger than that of older children and their parents. The mean disagreement is largest for pain and physical symptoms, which indicates less agreement on this scale than on the other scales. Another index quantifying the agreement between child and parent-reports shows the same significant findings;

Pearson correlation coefficients ranged from 0.26 to 0.51. Our results are in line with trends in the general population. Theunissen et al. (1998), found that children from the general population reported lower HRQol than their parents regarding physical functioning, motor functioning, autonomy, positive emotional functioning and cognitive functioning. The Pearson correlation coefficients on HRQol, in their study, ranged from 0.44 to 0.61. Verrips et al. (2000) also found that children reported lower HRQol on pain and physical symptoms than did their parents.

A possible explanation for these discrepancies in child-parent ratings may be that perceptions of HRQol and its meaning vary between individuals and within an individual over time as a response to many factors. Children differ from adults in their views about health, the cause and treatment of illness. They may lack the necessary language skills, the cognitive abilities to interpret questions, and are less able to judge their functioning in general. Their judgements may be heavily influenced by recent incidents (Vogels et al., 2000). Parent ratings of HRQol of their child may be biased by social desirability, and their own feelings and additional life stresses.

Previous research has shown that parents were more able to rate the child's HRQol in observable domains (physical functioning) in contrast to less visible domains (such as social or emotional functioning). Theunissen et al. (1998), however, found that children and parents were least likely to agree about physical functioning, for both health status and HRQol, compared with any other domain. In our study we found discrepancies between child-parent ratings on both observable (pain and physical symptoms, motor functioning and autonomy) and less observable (positive emotional functioning) domains of HRQol.

Our results indicate that, overall, our ConHD children had more negative views about their functioning than their parents. This finding is in line with trends in the general population. Considering our findings, we conclude that in medical settings both ConHD children themselves and their parents should be considered and used as important informants.

Limitations of the present study

The present sample of children with treated ConHD contains a selection of four diagnostic groups, within specific age ranges, and is therefore not completely representative for all ConHD anomalies.

The TACQOL is an adequate, generic HRQol instrument, applicable to a broad spectrum of children and adolescents. It doesn't provide, however, a disease-specific module for children with ConHD. Furthermore, no normative data are available from parents of children aged 12-15 years.

Implications

Since ConHD children, aged 8-11 years showed low HRQol long-term after invasive treatment, special attention should be given to screening and identifying children at risk in this age group. Wherever possible, information should be collected from multiple informants (child, both parents, teacher).

In future, a disease-specific, cross-culturally widely applicable HRQol instrument should be applied in clinical practice of children with treated ConHD, in addition to generic quality of life measures, in order to understand the impact of treated ConHD experienced by children and their parents.

3

Long-term intellectual functioning and school-related behavioural outcomes in children and adolescents after invasive treatment for congenital heart disease

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Abstract

Aims

To assess long-term intellectual functioning and school-related behavioural outcomes in a patient sample that underwent invasive treatment for congenital heart disease (ConHD) between 1990 and 1995.

Methods

The Wechsler Intelligence Scale for Children-Revised was used to measure intellectual functioning and the Teacher's Report Form to assess teacher-reported behavioural and emotional problems.

Results

Overall, patients have IQ-scores that fall within the normal range. The total sample of ConHD children (N=117, 7-16 years old), however, obtained significantly lower mean scores on Verbal IQ and Verbal Comprehension than reference children.

Compared to same-aged reference peers from the general population, 7-11-year-old ConHD children obtained significantly lower mean scores on Total IQ, Verbal IQ, Verbal Comprehension, and Perceptual Organization. In contrast, scores of 12-16-year-old ConHD children appeared to be significantly lower on Verbal Comprehension only and significantly higher on Performance IQ.

No significant differences were found in intellectual functioning between ConHD boys and girls, neither between different diagnostic groups.

The school-related behavioural and emotional adjustment of this sample of children with treated ConHD was encouraging.

Conclusion

Overall, this sample of recently treated ConHD children and especially children aged 7-11 years, showed poorer intellectual functioning on several areas. These findings deserve further attention.

Introduction

In the past two decades, diagnostic modalities, pre-operative medical management, surgical techniques, peri-operative care of congenital heart defects have improved substantially. The age at corrective (versus palliative) operation was significantly reduced for most lesions. As a result of these important advances, the pattern of survival of congenital heart disease (ConHD) has changed (Wren & O'Sullivan, 2001). With an increasing number of survivors, interest in neurodevelopmental outcome and quality of life has grown.

Several medical factors, such as deep hypothermic circulatory arrest, pre-, and peri-operative complications and the number of cardiac operations for an individual child, have been reported to put operated children at increased risk for later psychosocial adjustment problems (Utens et al., 1998). Subtle neurological deficits and global developmental lags have been observed in groups of patients operated for ConHD (Majnemer & Limperopoulos, 1999). It can be assumed that improved cardiological outcome will improve (long-term) psychosocial functioning.

A number of studies have been conducted with regard to the neurodevelopmental outcome of children with ConHD. Some of them were conducted only preoperatively, most studies were executed short-term after surgery, and only a few studies were done longitudinally (pre- and postoperatively with an interval between assessment). Several studies had methodological drawbacks, such as the restriction to only one cardiac diagnostic group, only cyanotic patients, patients operated upon with older techniques or small sample sizes (Aldén et al., 1998; Gomelsky et al., 1998; Jedlicka-Köhler et al., 1995; Kramer et al., 1989; Wright & Nolan, 1994). This hampers the possible generalization of findings. Furthermore, the focus of attention was on neurological abnormalities and intellectual dysfunction, whereas actual long-term schoolfunctioning has been relatively neglected. Studies in which both the long-term intellectual functioning and school-related behavioural outcomes are assessed in a recent patient sample, are lacking.

The present study is part of a follow-up study concerning the long-term medical and psychological outcome in children, adolescents and young adults who underwent invasive treatment for ConHD recently, that is between 1990 and 1995.

The aims of the present study were:

1. To compare the level of intellectual functioning in children and adolescents after invasive treatment for ConHD at a young age with that of a normative group.
2. To identify the role of sex, age, and cardiac diagnosis on the level of intellectual functioning.
3. To investigate the relation between behavioural and emotional problems as reported by teachers and intellectual functioning for this sample.

Methods

Wechsler Intelligence Scale for Children

The Wechsler Intelligence Scale for Children-Revised (WISC-R) (Wechsler, 1974) belongs to the most frequently used intelligence tests in clinical practice. The Dutch version of the WISC-R (Van der Steene et al., 1986) was used to assess intellectual functioning. The WISC-R consists of 12 subtests and yields three intelligence quotients (IQs). Firstly, Total IQ, which represents a global measure of the intellectual capacity. It is based on the scores on all subtests. Secondly, Verbal IQ, which gives an overall index for the child's capacity in verbal comprehension and verbal reasoning. It is based on the scores on the six verbal subtests: Information, Similarities, Arithmetic, Vocabulary, Comprehension, and Digit Span. And thirdly, Performance IQ, which is an estimate of perceptual organization skills, based on the scores of the six performance subtests namely: Picture Completion, Picture Arrangement, Block Design, Object Assembly, Coding, and Mazes. The psychometric properties of the WISC-R (reliability, validity) have been proven to be satisfactory (Van der Steene et al., 1986).

WISC profile analysis contributes to a better understanding of the intellectual capacities of a child by revealing strenghts and weaknesses in his or her profile (Kaufman & Reynolds, 1983). Profile analysis is executed by applying standardized procedures. It results in a description and interpretation of typical WISC patterns. Factor-analytic studies (Wechsler, 1974; Van der Steene et al., 1986) have revealed three factors underlying test performance. These are: 1) Verbal Comprehension, a factor that is formed by four verbal subtests representing verbal reasoning: Information, Similarities, Vocabulary, and Comprehension; 2) Perceptual Organization, which encompasses five performance subtests and represents perceptual organization: Picture Completion, Picture Arrangement, Block Design, Object Assembly, and Mazes; and 3) Freedom From Distractibility, a factor that is formed by three subtests, which refers to attention and concentration: Arithmetic, Digit Span, and Coding.

Teacher's Report Form

The Teacher's Report Form (TRF) (Achenbach, 1991c) is a standardised report on children's and adolescents' emotional and behavioural problems in the previous two months, as reported by teachers. The problem section consists of 120 problem items. Items are scored on a three-point scale: 0=not true; 1=somewhat or sometimes true; 2=very true or often true. The questionnaire consists of eight scales, that measure two broad problem areas: 'Internalising' problems reflect internal distress and 'Externalising' problems reflect conflicts with other people. 'Internalising' is measured by the scales Withdrawn, Somatic Complaints and Anxious/Depressed, and 'Externalising' by the scales Delinquent and Aggressive Behaviour. The syndrome scales Social Problems,

Thought Problems, and Attention Problems belong neither to the internalising nor the externalising group. A Total Problem score is computed by summing the individual item scores. Good reliability and validity for the Dutch version of the TRF has been reported (Verhulst et al., 1997a).

Inclusion and exclusion criteria

During the follow-up, which took place in 2003-2004, consecutive surviving patients of 4 diagnostic groups, who underwent their first invasive treatment for ConHD at least 7 years and 6 months ago (between 1 January 1990 and 1 January 1996) in the Erasmus University Medical Centre Rotterdam, and who were younger than 15 years at the time of the treatment, were eligible. The sample in this study encompassed the following cardiac diagnostic groups: surgical closure of atrial septal defect (ASD), surgical closure of ventricular septal defect (VSD), arterial switch operation of transposition of the great arteries (TGA) and balloon dilatation for pulmonary stenosis (PS). All operations were done with standard cardiopulmonary bypass. Patients with proven syndromes of mental retardation including Down syndrome were excluded.

Patient sample

The target population consisted of 246 consecutive surviving patients. At follow-up 40 patients were lost (23 moved abroad, 17 were untraceable). For the present study, 168 patients aged 7 to 16 years (upper age-range of the intelligence test) and their teachers were eligible. 38 Patients aged 17-28 years were not included since the instruments did not cover this age-range. 51 Children refused to participate (37 refused on practical and 14 on emotional grounds). The group non-responders consisted of 51% boys (n=26; mean age 11) and 49% girls (n=25; mean age 11). The final study sample consisted of the following subsamples of patients between 7-16 years and their teachers:

1. WISC-R patient sample.
   A total of 65 boys and 52 girls (n=117) provided usable WISC-R intelligence tests. The response rate of this subsample was 70%.
2. TRF teacher sample.
   Of the WISC-R patient sample, 6 parents didn't give informed consent to fill in the TRF and for 4 patients the TRF could not be filled in because they changed from (school and) teacher last 2 months. Furthermore, for 24 patients the TRF was not returned. Teachers of 45 male and 38 female patients (n=83) provided usable TRFs.

The numbers of patients in each cardiac diagnostic group of the WISC-R patient sample was: surgical closure of ASD n=26 (mean age 14.3 years, SD=1.8), surgical closure of VSD n=45 (mean age 11.9 years, SD=2.1), arterial switch operation of TGA n=32 (mean age 10.7 years, SD=1.4) and balloon dilatation for PS n=14 (mean age 13.2 years, SD=2.7).

Since in previous studies a correlation was found between socio-economic status (SES) of the family and child intelligence (Bloom et al., 1997; Forbess et al., 2002a, 2002b; Bellinger et al., 2003), SES was scored on a 9-point scale of parental occupation. Scores 1 to 3 corresponded with elementary and so-called 'lower' occupations (SES-level 1), 4 and 5 with so-called 'middle' occupations (SES-level 2), and 6 to 9 with so-called 'higher' and scientific occupations (SES-level 3) (Netherlands Central Bureau of Statistics, 1993). The percentages on each occupational level for families in the WISC-R patient sample was as follows: 28% SES-level 1 (n=33), 45% SES-level 2 (n=53), 27% SES-level 3 (n=31).

Reference groups

Data of the patient sample were compared with data derived from Dutch normative samples from the general population with comparable age-ranges.

1. WISC-R reference group.
   For the WISC-R Dutch age norms are available based on a normative sample of 1961 six- to 16-year-olds (Van der Steene et al., 1986). The population mean is 100 and the standard deviation 15. The mean for the twelve subtests is 10 and the standard deviation 3.
2. TRF reference group.
   For ages 7-16 years, the reference group consisted of 1239 children (648 boys and 591 girls) (Verhulst et al., 1997a).

Assessment procedure

The research protocol was approved by the Dutch Central Committee on Medical Research involving Human Subjects before the start of the study. All patients were traced and approached uniformly. After an information letter was received, patients were called for an appointment by the research assistant. Parents and/or patients signed an informed consent and returned it by mail, before participating in the study. The definite cardiac diagnosis was checked by a paediatric cardiologist (W.H.). During the visit in the Erasmus University Medical Centre, intellectual functioning of 7-16-year-old patients was assessed by a psychologist (A.S.). Parents were asked to hand the TRF plus informed consent to the teacher at school, who was most familiar with their child. Teachers filled in the questionnaire and returned it by mail in a prepaid envelope.

Statistical analyses

All analyses were performed with two-sided tests; $p<0.05$ was considered significant. One sample t-tests were used to test differences in group means between the patient sample and reference group on the WISC-R and TRF. For these analyses, Cohen's D's (Cohen, 1988) were computed to assess the magnitude of differences in mean scores between the patient sample and the reference group. According to Cohen's (Cohen, 1988)

criteria, a standardised difference of 0.20 can be considered as small, 0.50 as medium, and 0.80 as high.

To identify the role of sex (boys versus girls), age (7-11 versus 12-16 years) and cardiac diagnosis (ASD, VSD, TGA, PS) analyses of covariance, ANCOVAs were performed on all WISC-R scores. Because the variable socio-economic status (SES: 3 levels) showed a significant main effect in ANOVAs on all WISC scores it was applied as covariate. According to Cohen (Cohen, 1988), effects accounting for 1.0-5.9% of variance are considered small, 5.9-13.8% medium and >13.8% large.

To investigate the relation between intellectual functioning and school-related behavioural outcomes, Pearson correlations were performed on the WISC-R IQ-scores and the Total problems score of the TRF.

Results

Comparison of intellectual functioning of the total ConHD sample versus reference children

Table 1 shows the mean scores and standard deviations on the WISC-R for the ConHD patient sample by each sex. Two age categories (7-11 and 12-16 years) were formed based on the median split in the patient sample. Table 1 also shows significant differences and the corresponding effect sizes (Cohen's D). Compared with the reference group, the total sample of ConHD children obtained significantly lower scores on Verbal IQ and Verbal Comprehension. Verbal Comprehension appeared to be significantly lower both in ConHD boys and ConHD girls compared to same-sex peers from the general population. ConHD girls had a significant lower score on Verbal IQ than reference peers. Overall, mean WISC scores fell in the average range. Of the ConHD children 13% (n=15) had Total IQ scores below 85 and two of them below 70.

As to age groups, ConHD children, aged 7-11 years, obtained significantly lower mean scores on Total IQ, Verbal IQ, Verbal Comprehension, and Perceptual Organization than same-aged peers from the general population. For ConHD children, aged 12-16 years, two significant differences were found. Performance IQ appeared to be significantly higher and Verbal Comprehension appeared to be significantly lower compared to same-aged reference peers.

Table 2 shows the mean WISC-R subtest scores and standard deviations for the total ConHD sample. Scores on 10 of the 12 subtests were significantly different from the expected population mean score of 10. As to the verbal subtests, the total sample of ConHD children obtained significantly lower mean scores on Information, Vocabulary and Comprehension and a higher mean score on the subtest Similarities than reference peers. As to the performal subtests, ConHD children had lower mean scores on Picture

Table 1. Mean WISC-R scores, standard deviations and Cohen's D for the total ConHD sample

	Boys n=65		Girls n=52		7 - 11 years n=57		12 - 16 years n=60		Total sample n=117		
	Mean	**SD**	**Mean**	**SD**	**Mean**	**SD**	**Mean**	**SD**	**Mean**	**SD**	**Cohen's D**
Total IQ	98.2	14.1	98.7	13.4	95.6[1]	13.3	101.0	13.7	98.4	13.7	0.1
Verbal IQ	97.0	13.8	95.8[1]	11.6	95.2[1]	12.7	97.6	13.0	96.5[1]	12.9	0.2
Performance IQ	100.1	15.0	102.4	15.2	97.3[2]	14.5	104.7[1,2]	14.8	101.1	15.1	0.1
Verbal Comprehension	95.1[1]	13.0	96.2[1]	11.4	94.4[1]	11.8	96.7[1]	12.6	95.6[1]	12.3	0.3
Perceptual Organization	98.4	16.1	99.2	15.3	93.8[1,2]	15.6	103.5[2]	14.4	98.8	15.7	0.1
Freedom from Distractibility	102.7	16.9	102.1	13.2	101.7	15.9	103.2	14.9	102.4	15.3	0.2

Mean population score is 100; SD 15.

[1] Significant ($p<0.05$) difference between ConHD sample versus corresponding reference groups.

[2] Significant ($p<0.05$) difference between younger (7-11 years) versus older (12-16 years) ConHD patients.

Completion, Block Design, and Object Assembly. Compared with reference peers higher mean scores were found on the subtests Picture Arrangement, Coding and Mazes. No differences were found on the subtests Arithmetic and Digit Span. Although the mean scores on some subtests are below population means and others above, overall, ConHD children had a relative harmonious WISC profile and scored within the normal range.

Table 2. Mean WISC-R subtest scores and standard deviations for the ConHD sample

Subtests		ConHD n=117	
		Mean	SD
Verbal	Information	9.0*	2.7
	Similarities	10.8*	2.8
	Arithmetic	10.2	2.7
	Vocabulary	9.2*	2.6
	Comprehension	8.2*	2.2
	Digit Span	9.8	2.8
Performal	Picture Completion	8.7*	3.0
	Picture Arrangement	11.1*	2.9
	Block Design	9.4*	3.1
	Object Assembly	8.6*	3.5
	Coding	11.0*	3.1
	Mazes	11.9*	2.9

Mean population subtest score is 10; SD 3.

* Significant ($p<0.05$) difference between ConHD sample versus reference group.

Sex, age, and diagnosis within the patient sample

ConHD boys did not differ from ConHD girls on any of the WISC scores mentioned in Table 1. As to the separate subtests (Table 2), ConHD girls scored significantly lower on Arithmetic than ConHD boys. ConHD boys obtained significantly lower scores than ConHD girls on Comprehension and Coding.

ConHD children, aged 7-11 years, had significantly lower mean scores on Performance IQ and Perceptual Organization than ConHD children aged 12-16 years. Regarding separate subtests, 7-11-year-old ConHD children scored significantly lower on Block Design and Object Assembly than ConHD children aged 12-16 years.

Table 3 shows the mean scores and standard deviations on the WISC-R for different cardiac groups within the patient sample. Within the ConHD sample no significant differences for cardiac diagnosis on any of the WISC scores were found. Neither any difference was found between mean WISC scores of children treated by surgical intervention (ASD, VSD and TGA) and treated by catheter intervention (PS).

Table 3. Mean WISC-R scores and standard deviations for different cardiac diagnostic groups

	Cardiac diagnosis								
	ASD n=26		VSD n=45		TGA n=32		PS n=14		
	Mean	SD	Mean	SD	Mean	SD	Mean	SD	Main Effects
Total IQ	103.9	12.2	96.7	13.0	95.0	13.4	101.1	17.1	ns
Verbal IQ	99.5	11.4	97.0	12.8	93.4	13.7	95.9	13.6	ns
Performance IQ	108.1	14.2	97.4	13.3	98.0	13.6	107.3	19.8	ns
Verbal Comprehension	98.4	11.8	95.5	12.3	93.4	13.2	95.4	11.1	ns
Perceptual Organization	105.3	14.0	95.7	14.7	95.8	14.9	103.1	19.9	ns
Freedom from Distractibility	107.6	12.2	101.1	14.8	98.8	14.9	105.4	20.6	ns

Relationship between school-related behavioural outcomes and intellectual functioning

At follow-up 16 children (14%) of the total sample appeared to receive special education services and 39 (33%) had doubled in grade in the past. Table 4 shows the mean TRF scores and standard deviations for the ConHD patient sample and the reference group. Overall, no differences were found between teachers' reports regarding ConHD children's emotional and behavioural problems and those of the normative group, apart from one exception. Teachers of ConHD children reported significantly more Somatic complaints. Within this scale the item 'tired' is most frequently reported.

Pearson correlations were calculated between all WISC IQ- and factor scores and the Total problems score of the TRF. The Pearson correlation coefficients ranged from -0.23 to -0.31. All correlations were significant at a significance level of $p<0.05$. This means that ConHD children with poorer intellectual functioning showed more behavioural and emotional problems as reported by their teachers. According to Cohen (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 0.50 are considered large. Medium correlations were found between the Total IQ score and the Total problems score of the TRF, and on Freedom from Distractibility and the Total problems score of the TRF.

Table 4. Mean problem scores and standard deviations for TRF scales of the ConHD sample and reference group

	TRF			
	Mean problem scores			
	ConHD n=83		Reference n=1239	
Scales	**Mean**	**SD**	**Mean**	**SD**
Anxious/Depressed	4.1	5.2	3.6	4.3
Withdrawn	2.0	3.0	2.1	2.6
Somatic Complaints	1.0*	1.6	0.4	1.1
Social problems	2.9	3.5	2.2	3.3
Thought problems	0.6	1.5	0.4	0.9
Attention problems	7.7	7.0	6.8	6.9
Aggressive behaviour	5.4	7.0	4.7	7.3
Delinquent behaviour	0.7	1.3	0.8	1.6
Internalising	6.8	8.1	6.1	6.6
Externalising	6.1	7.8	5.4	8.4
Total problems	23.8	21.3	20.2	20.3

* Significant ($p<0.05$) difference.

Discussion

Intellectual functioning in the total patient sample compared to reference children

Although the mean WISC IQ- and factor scores are within the normal range, the results of the present study showed significantly lower scores regarding Verbal IQ and Verbal Comprehension for the total sample of ConHD children compared to the reference group. ConHD children, aged 7-11 years, had significantly poorer intellectual functioning than reference children on 4 areas: Total IQ, Verbal IQ, Verbal Comprehension, and Perceptual Organization. Scores of 12-16-year-old ConHD children were, overall, similar to those of reference peers, which is a favourable outcome. Only one score, namely Verbal Comprehension appeared to be lower in this age group. Overall, in our study we found that especially 7-11-year-old ConHD children showed poorer intellectual functioning.

In this study mean WISC IQ-scores were in the normal range, that is within 1 SD (±15) of the normative population mean of 100. This is in line with several previous studies, which showed that the IQ-scores of school age children were within the normal range (Forbess et al., 2002a, 2002b; Stavinoha et al., 2003; Visconti et al., 1999; Wray & Sensky, 2001). A main finding of our study was that poorer outcomes regarding Verbal IQ and Verbal Comprehension were found for the total sample of ConHD children. Previous studies also showed unfavourable verbal outcomes for certain cardiac defects (Bellinger et al., 2003; Wypij et al., 2003). In the Boston Circulatory Arrest Trial, assignment to total circulatory arrest was associated with lower Verbal IQ and Verbal Comprehension among children with TGA in combination with a VSD (Bellinger et al., 2003). Overall, results between studies are difficult to compare because of use of different diagnostic groups, use of different instruments, different ages at testing and follow-up periods, and surgical interventions done at different times at different centers by different surgical groups.

WISC analysis also showed that scores of our sample on profile factors and on all 12 subtests were within the normal range. Interestingly, we found a relative high score on Freedom from Distractibility, a factor that refers to attention and concentration. This favourable finding is in contrast with literature in which specific problem areas in attentional performance for subgroups of children with ConHD have been reported (O'Dougherty et al., 1988).

The present findings support previous outcomes of this sample, which showed lower outcomes on health-related quality of life regarding cognitive functioning, as reported by 8-15-year-old ConHD patients and their parents compared to reference groups (Spijkerboer et al., 2006).

Effect of sex, age and cardiac diagnosis within the patient sample

In this study the overall intellectual functioning of ConHD boys and ConHD girls was similar. As to age effects, we found in this sample that 7-11-year-old ConHD children

showed a poorer intellectual functioning on Performance IQ and Perceptual Organization than ConHD children aged 12-16 years.

No significant differences were found as to the intellectual functioning of the different cardiac diagnostic groups. This is in line with the study of Oates et al. (1995b). Neither any difference was found between intellectual functioning of children treated by surgical intervention and treated by catheter intervention. In a follow-up study, Visconti et al. (1999) found that children whose secundum atrial septal defect was closed with the use of a transcatheter device scored significantly higher on Total IQ, Performance IQ, and Perceptual Organization than children who had undergone open heart surgery. The group difference in Total IQ mainly reflected group differences in visual-motor and visual-spatial skills rather than in language-based skills. The study methods of Visconti et al. (1999), however, differed from ours, since they used only one diagnostic group and measured developmental outcome after surgical versus interventional closure of secundum atrial septal defect, whereas we assessed intellectual functioning in 4 diagnostic groups and the children with ASD in our sample all had undergone surgical closure.

Bellinger et al. (2003) found that children with TGA assessed at 8 years of age had lower scores on Total IQ, Performance IQ, Perceptual Organization, and Freedom from Distractibility than the expected population means. Our study showed that the Total IQ score and the verbal outcomes of children with TGA fell in the normal range, though all scores were lower than the expected mean of 100. Hövels-Gürich et al. (2002b) reported no significant differences in intellectual functioning in school-aged children who underwent in the past a neonatal arterial switch operation and normal healthy children.

School-related behavioural outcomes in relation with intellectual functioning

In previous studies, IQ was often assessed without measuring behavioural and emotional problems at school. Overall, in this study no differences were found in teachers' reports of school-related behavioural and emotional functioning for ConHD children versus reference children from the general population, apart from one exception. Only on Somatic Complaints, teachers of ConHD children reported more problems.

Our findings confirm those of Oates et al. (1994), who found that teachers of children, who had undergone cardiac surgery, did not report more emotional and behavioural difficulties than teachers of control children. Their study methods differed from ours: they compared teacher perceptions for a group children operated upon for ASD, VSD, TGA, and tetralogy of Fallot, 4 to 8 years previously, with those of a non-surgical control group. Our results are favourable, considering the finding of Karl et al. (2004). In their long-term follow-up, they compared patients who had undergone the arterial switch operation for TGA with best-friend control subjects on various aspects of school-related behaviour. Teachers of ConHD children reported more problems as to motor aspects of speech, language expression, or learning ability compared to teachers of control

children. Patients were reported to be more withdrawn, restless, or inattentive. Karl et al. (2004), however, used a different instrument (a self-designed list based on the TRF), included one diagnostic group and made comparisons with a control group, whereas we used normative reference data of a standardized and international well known questionnaire.

The item 'tired', from the Somatic Complaints factor, is the most frequently reported item by the teachers of our sample. This could reflect the teacher's awareness of the medical condition of the ConHD children, which may have lead to overreporting.

Investigating the relationship between intellectual functioning and teachers' reports of behavioural and emotional problems we found that ConHD children with poorer intellectual functioning showed more behavioural and emotional problems. As far as we know, no previous study has reported about this.

Limitations of the present study

The present sample of children with ConHD contains four diagnostic groups, within specific age ranges, and is therefore not completely representative for all ConHD anomalies. Though our response rate was satisfactory, intellectual functioning and school-related behavioural and emotional outcomes could not be assessed in all eligible patients. To what extent this may have influenced our data is unknown.

Implications

Overall, our patients have IQ scores that, on average, fall within the normal range at time of follow-up (at least 8 years after invasive treatment for ConHD). The school-related behavioural and emotional adjustment of this sample is encouraging. Teacher reports of behavioural and emotional problems in these ConHD children were similar to teachers' reports of reference peers. These findings, however, do not preclude patients to be at risk. In our sample 13% of the children were mentally retarded or had borderline intellectual functioning. The number of patients evaluated who received special education services (14%) and had doubled in grade in the past (33%) reveals problems in a substantial portion of this population.

Our results indicate that, overall, the most prominent deficits lie in the domains of Verbal IQ and Verbal Comprehension. Since 7-11-year-old ConHD children showed lower intellectual functioning on several areas long-term after invasive treatment, special attention should be given to screening and identifying children at risk in this age group. Health care providers/professionals need to be aware which patients are vulnerable for intellectual dysfunction and difficulties in schoolfunctioning. Therefore, we recommend structural (neuro)developmental follow-up of children who have undergone invasive treatment for ConHD.

4

Behavioural and emotional problems in children and adolescents long-term after invasive treatment for congenital heart disease

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Abstract

Aims

To assess the occurrence of a wide range of behavioural and emotional problems long-term after invasive treatment for congenital heart disease (ConHD) in infancy and childhood.

Methods

Parents of 125 ConHD children, aged 7-17, completed the Child Behavior Checklist and 85, 11-17-year-old, ConHD children completed the Youth Self-Report.

Results

Parents of ConHD children and adolescents evaluated the patients' behavioural problems as significantly more unfavourable than parents of the reference group. Parents reported significantly higher problems scores for the scales Somatic Complaints, Social Problems, Attention Problems, Internalising and Total Problems compared to the reference group. In contrast, reports of patients were comparable to those of reference peers. No differences were found on the self-reports between problem scores for different cardiac diagnostic groups.
Discrepancies between self- and parent-reports were found, indicating that more problems were reported by ConHD patients themselves than by their parents.

Conclusion

Overall, parents of ConHD patients reported higher levels of behavioural and emotional problems compared to the reference group whereas patients themselves reported no long-term behavioural impairment. Younger ConHD patients deserve special attention. Assessing behavioural and emotional problems in ConHD patients can be helpful to detect children at risk for developing psychopathology.

Introduction

Over the past decades, advances in diagnostic and surgical techniques and medical treatment of congenital heart disease (ConHD) have gradually evolved and have significantly improved long-term survival of patients with ConHD (Eskedal et al., 2005; Boneva et al., 2001).

Children with (chronic) physical health problems are vulnerable for behavioural and emotional problems (Lavigne & Faier-Routman, 1992; Wallander & Varni, 1998). Research has shown that children and adolescents with ConHD exhibit more behavioural and emotional problems than children from the general population (Fredriksen et al., 2004; Hövels-Gürich et al., 2002; Janus & Goldberg, 1995; Utens et al., 1993), irrespective of the severity of the disease (Fredriksen et al., 2004; Utens et al., 1993). In a follow-up study conducted about 15 years ago, Utens et al. (1993) found significantly more behavioural and emotional problems in children and adolescents with ConHD (27%) compared to same-aged peers from reference groups (10%) at least 9 years after cardiac surgery. Since 1980, the most recent year of cardiac surgery included in this ConHD study, many aspects of medical care for ConHD have changed. The age at corrective (versus palliative) operation has been reduced significantly for most conditions. New techniques, including catheter intervention, have been introduced for other lesions. These changes are generally believed to have improved cardiological outcome in most types of ConHD. It may be hypothesised that the presumed improvement in cardiological outcome has improved (long-term) behavioural and emotional adjustment of the children treated upon. Therefore, this study's aim was to systematically investigate the level of behavioural and emotional problems of more recently treated ConHD children.

The present study is part of a follow-up study concerning the long-term medical and psychological outcomes in children, adolescents and young adults who underwent invasive treatment for ConHD between 1990 and 1995.

The aims of the present study were:

1. To compare the level of behavioural and emotional problems in children and adolescents who underwent invasive treatment for ConHD at a young age with that of same-aged reference groups.
2. To identify the role of sex, age and cardiac diagnosis on the level of behavioural and emotional problems.
3. To determine the agreement between child and parent ratings within this sample.

Methods

Instruments

The Child Behavior Checklist (CBCL) (Achenbach, 1991a) was used to obtain standardized parents' reports of behavioural and emotional problems in children aged 7-17 years. The problem section consists of 120 problem items. Parents rate their child's behaviour during the preceding six months on a 3-point scale (0 = not true; 1 = somewhat or sometimes true; 2 = very true or often true).

For 11-17-year-olds the Youth Self-Report (YSR) (Achenbach, 1991b) was used to obtain adolescents' self-reports. The YSR was modeled after the CBCL and has the same format, except that items are worded in the first person. Good validity and reliability of the CBCL and YSR have been established (Achenbach, 1991a, 1991b; Achenbach & Rescorla, 2001) and were confirmed for the Dutch translations (Verhulst et al., 1996, 1997b).

Both questionnaires consist of eight specific syndrome scales and two broad problem areas Internalising and Externalising. Internalising problems reflect internal distress and Externalising problems reflect conflicts with other people. The Internalising scale consists of the syndrome scales Withdrawn, Somatic Complaints and Anxious/Depressed, whereas the Externalising scale consists of Rule-Breaking and Aggressive Behaviour. The syndrome scales Social Problems, Thought Problems, and Attention Problems belong neither to the Internalising nor the Externalising group. A Total Problems score can be obtained by summing the scores on all individual problem items. A higher score indicates a higher level of problems.

Inclusion and exclusion criteria

During the follow-up, which took place in 2003-2004, consecutive surviving patients of 4 diagnostic groups, who underwent their first invasive treatment for ConHD at least 7 years and 6 months ago (between 1 January 1990 and 1 January 1996) in the Erasmus University Medical Centre Rotterdam, and who were younger than 15 years at the time of the treatment, were eligible. The sample in this study encompassed the following cardiac diagnostic groups: surgical closure of atrial septal defect (ASD), surgical closure of ventricular septal defect (VSD), arterial switch operation of transposition of the great arteries (TGA) and balloon dilatation for pulmonary stenosis (PS). Patients with proven syndromes of mental retardation including Down syndrome were excluded.

Patient sample

The target population consisted of 246 consecutive surviving patients. At follow-up 40 patients were lost (23 moved abroad, 17 were untraceable). Of the remaining 206 patients, 35 patients aged 18-28 years were not included since the instruments did not cover this age-range. 171 Patients were aged 7-17 years (age-range of the parent-report,

CBCL) at follow-up. 115 Patients were 11-17 years (age-range of the YSR). From the CBCL sample parents of 46 patients refused to participate for practical or emotional reasons. Of these 46, 24 patients were aged 7-12 and 22 patients 13-17 years. The group non-responders of the YSR patient sample consisted of 14 boys (mean age 13) and 16 girls (mean age 13).

The subsamples of the present study partly overlapped since the CBCL and YSR partly covered the same age-ranges. The final study sample consisted of the following subsamples of patients and their parents:

1. CBCL patient sample.
   Parents of the 125 children aged 7-17 years completed the CBCL. One questionnaire was not usable because of incomplete information. The final CBCL sample consisted of parents of 66 male and 58 female patients (n=124). The response rate of this subsample was 73%.
2. YSR patient sample.
   85 Participating ConHD children aged 11-17 years completed the YSR. One questionnaire was not usable because of incomplete information. The final YSR patient sample consisted of 44 boys and 40 girls (n=84). The response rate of this subsample was 73%.

Reference groups

Data of the CBCL and YSR patient samples were compared with data derived from Dutch normative samples from the general population, with comparable age-ranges assessed with the same instruments (Tick et al., submitted).

1. CBCL reference group.
   The CBCL reference group consisted of parents of 713 boys and 726 girls, aged 7-17 years.
2. YSR reference group.
   The YSR reference group consisted of 349 boys and 382 girls between 11 and 17 years.

Socio-economic status (SES) was scored on a 9-point scale of parental occupation. Scores 1 to 3 corresponded with elementary and so-called 'lower' occupations (SES-level 1), 4 and 5 with so-called 'middle' occupations (SES-level 2), and 6 to 9 with so-called 'higher' and scientific occupations (SES-level 3) (Netherlands Central Bureau of Statistics, 1993). The percentages on each occupational level for the CBCL and YSR patient samples respectively were as follows: SES-level 1: 27.1 and 28.6%; SES-level 2: 48.3 and 42.9%; SES-level 3: 24.6 and 28.6%. The results of the CBCL and YSR patient samples were different because the samples consisted of persons with different ages. The percentages for the CBCL and YSR reference groups were respectively: SES-level 1: 24.1 and 22.7%; SES-level

2: 41.3 and 39.2%; SES-level 3: 34.6 and 38.1%. There were no significant differences in the SES distribution between the CBCL and YSR patient samples and reference groups (CBCL: $\chi^2 = 5.28$, df = 2, ns; YSR: $\chi^2 = 3.56$, df = 2, ns).

Assessment procedure

The research protocol was approved by the Dutch Central Committee on Medical Research involving Human Subjects before the start of the study. All patients were traced and approached uniformly. After an information letter was received, patients were called for an appointment by the research assistant. Parents and/or patients signed an informed consent and returned it by mail, before participating in the study. The definite cardiac diagnosis was checked by a paediatric cardiologist (W.H.). During the psychological investigation in the Erasmus University Medical Centre, patients aged 11-17 years completed the YSR in the presence of a psychologist (A.S.) Parents of patients, aged 7-17 years, completed the CBCL independently from their child in the waiting room.

Statistical analyses

Differences in proportions scoring in the deviant range between patient samples versus reference groups were analysed by Binomial Testing. Differences in mean scores of the patient samples versus reference groups were assessed with analyses of variance (ANOVAs). To investigate agreement between different informants (parents and children) respectively to test differences between informants, MANOVAs were performed on the CBCL and YSR scales of 11-17-year-old patients with both a parent- and child-report completed.

Results

Proportions of problem children

Table 1 shows the proportions of children in the ConHD patient sample and reference group who scored in the deviant range of the CBCL (parent-reports) or the YSR (self-reports). The 90th percentiles of the cumulative frequency distributions of the CBCL and YSR Total Problem scores obtained for the reference group were chosen as the cut-offs to distinguish problem from non-problem children. Due to gaps between rank ordered scores, the percentages from the reference group scoring above the 90th percentile were not exactly 10%. To assess age effects, two categories were formed based on the median split of the patient samples. In the CBCL patient sample for both sexes, two age groups, 7-12 and 13-17 years, were formed. The two age categories in the YSR patient sample were as follows: 11-12 and 13-17 years.

The percentage of patients who scored in the deviant range for the total CBCL sample (16.9) was significantly greater than that in the total CBCL reference group (10.2). A

significant difference was found between male patients (21.2%) versus reference boys (9.5%) scoring in the deviant range according to parent-reports. No significant differences were found in the percentages of patients who scored in the deviant range between the YSR patient sample and the YSR reference group.

Mean problem scores

Table 1 also shows the mean Total Problem scores on the CBCL and YSR by each sex and both age categories for the patient samples and the reference groups. To test differences in mean Total Problem scores and mean scale scores ANOVAs were computed in a group (patient sample versus reference group) x sex (boys versus girls) x age (CBCL: 7-12 versus 13-17 years; YSR: 11-12 versus 13-17 years) factorial design. Socio-economic status wasn't applied as covariate since it did not show significant main effects on the mean CBCL and YSR Total Problem scores in ANOVAs.

Table 2 shows the mean scale scores for the CBCL and YSR patient sample and reference group for the eight scales, Internalising and Externalising and Total Problem, as well as the ANOVA results. 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 (Cohen, 1988), effects accounting for 1.0-5.9% of variance are considered small, 5.9-13.8% medium and >13.8% large.

On the CBCL scales Somatic Complaints, Social Problems, Attention Problems, Internalising and Total Problems, parents of ConHD children reported significantly more problems than parents of children in the reference group. According to Cohen's criteria (Cohen, 1988) the group effects were small. No significant two-way interactions were found.

On the YSR a small, though significant group effect for Rule-Breaking Behaviour indicated that ConHD children reported these problems significantly less often than children in the reference group. Significant group x age interaction effects were found on the YSR for Anxious/Depressed, Thought Problems, Attention Problems, Rule-Breaking Behaviour, Aggressive Behaviour, Externalising, and Total Problems. All effects represented larger differences for the younger (11-12 years) versus older (aged 13-17 years) patients in the ConHD sample compared to those in the reference group, with the older ConHD patients scoring more favourably than the younger ConHD patients, whereas in the reference group the younger children scored more favourably than the older children.

For YSR Somatic Complaints, a significant group x sex interaction effect was found. The effect represented larger differences for boys verus girls in the reference group compared to the ConHD patient sample, with boys scoring more favourably than girls in both the reference group and the patient sample.

Table 2. Mean problem scores for CBCL and YSR scales for the ConHD samples and reference groups, and percentages of variance accounting for the difference between the ConHD and reference group in ANOVAs

Table 1. Percentages of the ConHD samples and reference groups scoring in the CBCL and YSR deviant range

	CBCL						YSR				
	Mean Total Problems scores		% > Cut-off[a]		Binomial Test *P*		Mean Total Problems scores		% > Cut-off[a]		Binomial Test *P*
	ConHD (n)	Reference (n)	ConHD	Reference			ConHD (n)	Reference (n)	ConHD	Reference	
Boys	31.7 (66)	25.9 (713)	21.2	9.5	0.005	**Boys**	33.3 (44)	31.6 (349)	9.1	9.7	ns
7 - 12 yr[b]	32.3 (42)	27.9 (400)	21.4	10.0	0.021	11 - 12 yr[b]	39.6 (21)	33.1 (102)	19.0	9.8	ns
13 - 17 yr[b]	30.5 (24)	23.3 (313)	20.8	10.5	ns	13 - 17 yr[b]	27.5 (23)	30.9 (247)	0	10.1	ns
Girls	28.3 (58)	25.1 (726)	12.1	9.5	ns	**Girls**	35.6 (40)	38.2 (382)	15.0	10.2	ns
7 - 12 yr[b]	30.5 (28)	24.2 (405)	14.3	10.1	ns	11 - 12 yr[b]	42.0 (13)	35.5 (98)	23.1	9.2	ns
13 - 17 yr[b]	26.2 (30)	26.2 (321)	10.0	10.3	ns	13 - 17 yr[b]	32.4 (27)	39.2 (284)	11.1	9.9	ns
Total sample	30.1 (124)	25.5 (1439)	16.9	10.2	0.011	**Total sample**	34.4 (84)	35.1 (731)	11.9	10.0	ns

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

[b] Age categories based on median split of patient samples.

	CBCL			YSR		
	Mean problem scores			Mean problem scores		
Scales	ConHD (n=124)	Reference (n=1439)	Group* (%)	ConHD (n=84)	Reference (n=731)	Group* (%)
Anxious/Depressed	3.6	3.2		3.9	4.2	
Withdrawn/Depressed	2.6	2.2		2.7	3.0	
Somatic Complaints	2.1	1.6	0.4	3.1	3.0	
Social Problems	3.4	2.3	1.5	3.6	3.3	
Thought Problems	2.4	2.0		3.3	3.1	
Attention Problems	5.0	4.0	0.7	5.1	5.1	
Rule-Breaking Behaviour	1.8	2.1		3.1	3.9	0.6
Aggressive Behaviour	5.4	4.7		5.3	5.3	
Internalising	8.3	7.0	0.3	9.7	10.3	
Externalising	7.2	6.7		8.4	9.2	
Total Problems	30.0	25.5	0.5	34.4	35.1	

* Percentage of variance accounted for by significant group effect

Diagnostic groups

Table 3 shows the mean Total Problem scores and mean scale scores of the different cardiac groups within the CBCL and YSR samples. Numbers of patients belonging to each cardiac diagnostic group of the CBCL sample were: surgical closure of ASD n=29 (mean age 14.0 years), surgical closure of VSD n=50 (mean age 11.3 years), arterial switch operation of TGA n=32 (mean age 10.4 years) and balloon dilatation for PS n=13 (mean age 12.5 years). The diagnostic groups in the YSR patient sample were as follows: ASD n=27 (mean age 14.4 years), VSD n=33 (mean age 12.4 years), TGA n=16 (mean age 11.5 years) and PS n=8 (mean age 13.8 years).

To test differences in mean problem scores across the diagnostic groups, ANOVAs were computed with cardiac diagnosis, sex and age as independent variables. Two main effects for diagnosis were found for CBCL Social Problems and CBCL Externalising ($p<0.05$). Post hoc test of least squared differences revealed that parents of children with VSD reported significantly higher scores on Social Problems and Externalising than parents of children with an ASD and PS.

In the YSR sample, cardiac diagnosis did not have a main effect on any of the scales, nor on the Total Problem score. No significant two- or three way interactions were found.

Different informants

To test effects of different informants (parents versus children) MANOVAs with informant as the within factor were performed on the scores of 11-17-year-old ConHD patients and the reference group with both a CBCL and YSR completed (n=83 respectively n=730).

Table 3. Mean problem scores of CBCL and YSR for different cardiac diagnostic groups

	CBCL					YSR				
Scales	**ASD (n=29)**	**VSD (n=50)**	**TGA (n=32)**	**PS (n=13)**	**Main effects**	**ASD (n=27)**	**VSD (n=33)**	**TGA (n=16)**	**PS (n=8)**	**Main effects**
Anxious/Depressed	3.2	4.1	3.5	3.2	ns	3.4	4.1	4.4	3.9	ns
Withdrawn/Depressed	2.6	2.7	1.9	3.9	ns	2.7	2.6	2.4	3.5	ns
Somatic Complaints	2.6	2.2	1.6	2.0	ns	2.9	2.9	3.9	2.9	ns
Social Problems	2.2	4.4	3.3	2.4	<0.05	3.3	3.5	4.4	3.0	ns
Thought Problems	1.7	3.0	2.6	1.6	ns	2.6	3.9	3.8	3.0	ns
Attention Problems	4.1	5.6	5.3	4.1	ns	5.1	4.3	6.3	5.4	ns
Rule-Breaking Behaviour	1.5	2.5	1.3	1.2	ns	2.7	3.5	3.6	2.4	ns
Aggressive Behaviour	3.7	6.5	5.7	3.8	ns	4.7	5.3	6.9	4.1	ns
Internalising	8.4	9.0	7.0	9.2	ns	9.0	9.5	10.6	10.3	ns
Externalising	5.2	9.0	7.1	5.0	<0.05	7.3	8.8	10.5	6.5	ns
Total Problems	24.9	35.1	29.0	24.9	ns	32.1	34.0	39.9	32.4	ns

Abbreviations used are: ASD = Atrial Septal Defect, VSD = Ventricular Septal Defect, TGA = Transposition of the Great Arteries, PS = Pulmonary Stenosis.

Significant informant x group interaction effects were found for Withdrawn/Depressed, Social Problems, Attention Problems, and Total Problems (all $p<0.05$), indicating that on these scales more problems were reported by children themselves than by their parents. All effects represented larger differences between children and parents in the reference group compared to the ConHD sample.

Discussion

Level of behavioural and emotional problems in the patient sample compared to the reference group

The present study's results showed that according to parents' reports of problem behaviours a significant proportion of ConHD children scored in the deviant range (16.9%) compared to the reference group (10.2%). The proportion of ConHD boys scoring in the deviant range according to parents was significantly greater than that in the reference sample. According to patients themselves (YSR sample) the proportion of children and adolescents scoring in the deviant range, was comparable to that in the reference group.

Parent-reports indicated significantly more total problems for ConHD children and adolescents and significantly more somatic complaints, social, attention, and internalising problems than for peers in the reference group. In contrast, reports of 11-17-year-old ConHD patients themselves were comparable or even better to those of reference peers, which is a favourable outcome. Compared to same-aged reference peers, ConHD children and adolescents reported significantly less problems on Rule-Breaking Behaviour.

In contrast to our present favourable findings regarding the self-reports of the 11-17-year-old ConHD children, Utens et al. (1993) found in their sample, treated for ConHD between 1968 and 1980, significantly more problems on all scales of the YSR compared to same-aged reference peers. Our findings are in line with those of previous studies, reporting elevated levels of behavioural and emotional problems in ConHD children and adolescents according to parents' reports (Janus & Goldberg, 1995; Utens et al., 1993; Oates et al., 1994). Our unfavourable outcome regarding the elevated proportion of ConHD boys scoring in the deviant range according parent-reports resembles that of Fredriksen et al. (2004). They found that parents of 11-16-year-old ConHD boys reported significantly higher Total Problem scores on the CBCL than a reference group. The study methods of Fredriksen et al. (2004), however, differed from ours, since patients with other ages, other types of cardiac diagnostic groups and surgical interventions were included. A strenght of our study is that we used four homogeneous cardiac diagnostic groups treated recently.

The higher CBCL Internalising score in comparison to the norm could be explained by Somatic Complaints. This finding could indicate that parents of ConHD children, due to alertness to illness related factors and bodily symptoms, have an attitude to report these symptoms more often for their children. It might also be explained by actual somatic problems in the ConHD patient sample. Previous studies reported that children with chronic illnesses are more likely to suffer from internalising problems (Fredriksen et al., 2004; Utens et al., 1993; Oates et al., 1994; Gupta et al., 2001) and are likely to experience social competence problems. Parents in our study reported more social and attention problems. From literature it is known that for ConHD children impairments in physical capacity are associated with difficulties in psychosocial functioning (Fredriksen et al., 2004; Bjørnstad et al., 1995; Spurkland et al., 1993). It can be speculated that parents reported more social problems for their ConHD children due to parental perceptions of reduced physical ability in their children interfering with peer relationships. The unfavourable finding regarding attention problems is in contrast with previous outcomes for this sample regarding the intellectual and schoolfunctioning as assessed with an IQ-test (Van der Steene et al., 1986) and the Teacher's Report Form (Achenbach, 1991c), which showed no specific problem area in attentional performance (Spijkerboer et al., submitted a).

In contrast to the parent-reports, the self-reports of the 11-17-year-old ConHD children showed favourable outcomes. Whether this is an underestimation of behavioural and emotional problems by patients themselves, due to social desirability or denial or a reflection of 'the truth' remains unclear. Previously we found a similar positive trend: overall ConHD adolescents (aged 12-15 years) in this sample reported a comparable level of health-related quality of life as same-aged reference peers (Spijkerboer et al., 2006).

Effect of sex, age and cardiac diagnosis on the level of behavioural and emotional problems

In this study no significant group x sex effects were found, except for the self-report Somatic Complaints; boys scored more favourably than girls in both the reference group and patient sample, with larger differences between sexes in the reference group. Further, no significant group x age effects were found on the parent-reports. For the self-reports, however, compared to the reference group, greater differences for several scales between younger (11-12 years) versus older (13-17 years) patients were observed, with older patients scoring more favourably while in the reference group the reversed situation was observed.

In this study, two significant main effects for cardiac diagnosis were found for the parent-reports of Social Problems and Externalising, indicating more problems in children with VSD versus ASD and PS. In contrast, our results showed no differences between the

self-reports on emotional and behavioural problems of different cardiac diagnoses. This is in line with previous studies (Fredriksen et al., 2004; Utens et al., 1993), which did not show any relationship between the type or severity of the cardiac defect and the level of behavioural and emotional problems in children and adolescents.

Agreement between child and parent rating

In this study significant informant x group effects were found, indicating that more problems were reported by children themselves than by their parents. All effects represented larger differences between child- and parent-reports in the reference group compared to the ConHD sample.

Previous outcomes in this sample showed a similar pattern: ConHD children and adolescents reported a poorer quality of life than did their parents about them (Spijkerboer et al., 2006). Considering our present and previous findings, we conclude that in the assessment of behavioural and emotional problems, information needs to be collected from both parents and children as well to identify children at risk.

Implications

Remarkably, despite advances in diagnostic and surgical techniques and medical treatment of ConHD resulting in improved cardiological outcomes, parents of ConHD children and adolescents evaluated the patients' behavioural problems as significantly more unfavourable than parents of the reference group. Since discrepancies between child and parent ratings of the level of behavioural and emotional problems were found, it is important to use both ConHD children themselves and their parents to judge their behaviour. Younger ConHD children showed more behavioural and emotional problems long-term after invasive treatment compared to older ConHD patients and same-aged reference peers, special attention should therefore be given to screening these problems at young age. This can reduce the risk for developing psychopathology.

5

A historical comparison of long-term behavioural and emotional outcomes in children and adolescents after invasive treatment for congenital heart disease

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Submitted for publication

Abstract

Aims

Children with congenital heart disease (ConHD) are known to be vulnerable to behavioural and emotional problems. In this study a historical comparison is made between the level of behavioural and emotional problems in a sample of children with ConHD treated recently, versus a comparable historical sample operated upon before 1980 in the same institute. The hypothesis was that improvements in medical care would result in more favourable behavioural and emotional outcomes for children with ConHD treated recently, that is between 1990 and 1995, compared to same-aged patients operated before 1980.

Methods

To assess behavioural and emotional problems the Child Behavior Checklist (parent-report) and Youth Self-Report were used. The historical samples (n=98 respectively n=123) and recent samples (n=90 respectively n=84) consisted of four diagnostic groups.

Results

Parents and children from the recent ConHD sample reported fairly similar levels of behavioural and emotional problems compared to parents and children in the historical ConHD sample.

Conclusion

Despite evident improvements in diagnostic and surgical techniques and medical treatment of ConHD over the past decades, virtually no changes were found in levels of problem behaviour of the recent patient sample compared to the historical patient sample, who both underwent invasive treatment for ConHD.

Introduction

Long-term survival of patients with congenital heart disease (ConHD) has improved significantly (Eskedal et al., 2005; Sparacino, 1994; Wren & O'Sullivan, 2001). This may be accompanied by increased long-term morbidity and impaired emotional adjustment. Research has shown that children with ConHD are at higher risk for behavioural and emotional problems than reference peers, irrespective of the severity of their ConHD (Fredriksen et al., 2004; Hövels-Gürich et al., 2002a; Janus & Goldberg, 1995; Utens et al., 1993). The last decades many aspects of diagnostic, surgical and medical treatment for ConHD have been improved.

In the present study, a historical comparison is made between the level of behavioural and emotional problems of a sample of children and adolescents with ConHD treated recently, that is between 1990 and 1995 (Spijkerboer et al., submitted b), and that of a historical sample of children and adolescents operated for ConHD between 1968 and 1980 (Utens et al., 1993). Both samples were treated in the Erasmus University Medical Centre Rotterdam. Behavioural and emotional problems are assessed by using well-known parent- and self-reports, commonly used with pediatric populations.

We hypothesize that the sample of children and adolescents with ConHD treated recently show better outcomes regarding their behavioural and emotional adjustment compared to the historical sample operated for ConHD between 1968 and 1980. So far, no previous study has been executed to make a historical comparison between the long-term behavioural and emotional outcomes of a recent sample versus a historical sample of children and adolescents treated for ConHD.

Methods

Child Behavior Checklist and Youth Self-Report

The Child Behavior Checklist (CBCL) (Achenbach, 1991a) and the Youth Self-Report (YSR) (Achenbach, 1991b) were used to obtain respectively standardized parents' and self-reports of children's behavioural and emotional problems over the preceding six months. The YSR is modelled after the parent-report except that the child-form is worded in the first person.

The questionnaires consist of eight specific syndrome scales, and two broad problem areas Internalising and Externalising. Internalising problems reflect internal distress and Externalising problems reflect conflicts with other people. The Internalising scale consists of the syndrome scales Withdrawn, Somatic Complaints and Anxious/Depressed, whereas the Externalising scale consists of Rule-Breaking and Aggressive Behaviour. The syndrome scales Social Problems, Thought Problems, and Attention Problems belong

neither to the internalising nor the externalising group. A Total Problems score can be obtained by summing the scores on all individual problem items. A higher score indicates a higher level of problems. Good validity and reliability have been established (Achenbach, 1991a, 1991b; Achenbach & Rescorla, 2001) and were confirmed for the Dutch version (Verhulst et al., 1996, 1997b).

Total study population

Both the recent and historical sample consisted of consecutive surviving patients of 4 diagnostic groups (atrial septal defect: ASD; ventricular septal defect: VSD; transposition of the great arteries: TGA; and pulmonary stenosis: PS), who underwent their first invasive treatment for ConHD in the Erasmus University Medical Centre Rotterdam, and who were younger than 15 years at the time of the treatment. In both samples, patients with proven syndromes of mental retardation including Down syndrome were excluded. Due to the age-ranges of instruments used in both samples, parents of 10-15-year-old patients (CBCL) and 11-17-year-old patients (YSR) from both samples were investigated.

Recent ConHD sample

The 4 cardiac diagnostic groups in the recent ConHD sample were treated as follows: surgical closure of ASD, surgical closure of VSD, arterial switch operation of TGA with or without VSD and balloon dilatation for PS. All operations were done between 1990 and 1995 with standard cardiopulmonary bypass.

The target population consisted of 246 consecutive surviving patients. At follow-up 40 patients were lost (23 moved abroad, 17 were untraceable). Of the remaining 206 patients, aged 7-28 years, 115 patients were 11-17 years (age-range of the YSR). The group non-responders of the YSR patient sample consisted of 14 boys (mean age 13) and 16 girls (mean age 13). 120 Patients were 10-15 years (age-range of the CBCL used in this study). 29 Parents from the CBCL patient sample refused to participate for practical or emotional reasons.

YSR patient sample:

From the 115 eligible patients aged 11-17 years, 85 patients completed the YSR. One questionnaire was not usable because of incomplete information. The final YSR patient sample consisted of 44 boys and 40 girls (n=84). The response rate was 73%.

CBCL patient sample:

From the 120 eligible 10-15-year-old patients, parents of 91 patients completed the CBCL. One questionnaire was not usable because of incomplete information. The final CBCL sample consisted of parents of 45 male and 45 female patients (n=90). The response rate was 75%.

Historical ConHD sample

The historical ConHD sample was selected from a study population of all consecutive patients who underwent their first open heart surgery for ConHD between 1968 and 1980. All patients in this sample received surgical treatment. The TGA group was operated using the Mustard technique. PS patients were operated upon using inflow occlusion or with the use of cardiopulmonary bypass. For details of the initial data collection, see Utens et al. (1993). To compare the levels of behavioural and emotional problems between the recent and historical sample we selected patients with the same 4 cardiac diagnoses. Thus, 98 parent-reports (CBCL) of 10-15-year-old patients and 123 self-reports (YSR) of 11-17-year-old children were selected.

Assessment procedure

For both the historical and recent sample the research protocol was approved by the ethics committee review board before the start of the study. In both studies, all patients were traced and approached uniformly. After an information letter was received, patients were called for an appointment by the research assistant. Before participating in the study, parents and/or patients signed an informed consent and returned it by mail. The definite cardiac diagnosis was checked by a paediatric cardiologist. During the psychological investigation in the Erasmus University Medical Centre, patients aged 11-17 years completed the YSR in the presence of a psychologist. Parents of patients completed the CBCL independently from their child in the waiting room. Data collection took place in the same standardized way from 1989 and 1991 (historical sample) and from 2003 and 2005 (recent sample).

Statistical analysis

Pearson Chi-square tests were used to test differences in distributions of sex and cardiac diagnosis, and t-tests were used to test differences in mean age at follow-up of patients in the recent and historical sample for both the CBCL and YSR. Differences in mean scores between the recent CBCL and YSR sample versus respectively the historical CBCL and YSR sample were assessed with analyses of (co)variance (AN(C)OVAs). Items 2, 4, 5, 28, 78, and 99 for both the CBCL and YSR were excluded from analysis, since these items did not overlap in the questionnaires completed by parents and patients in the historical sample versus those completed by parents and patients in the recent sample. The criteria by Cohen (Cohen, 1988) were applied to categorize effect sizes for ANOVAs. Effects accounting for 1.0-5.9% of variance are considered small, 5.9-13.8% medium and >13.8% large.

Results

Table 1 shows the patient characteristics of both the recent and the historical ConHD samples. No significant differences were found in the distribution of sex and mean age at follow-up between the recent and historical sample for the CBCL. For the YSR a significant difference in mean age at follow-up was found between the recent and the historical sample, with the historical sample containing patients on average about 1.5 year older at follow-up ($p<0.001$). No significant difference as to sex distribution was found for the YSR samples. Significant differences in the distribution of cardiac diagnosis between the recent and historical sample were found for both the CBCL and the YSR samples ($\chi^2 = 11.4$, $p=0.010$ respectively $\chi^2 = 13.1$, $p=0.004$).

Table 1. Patient characteristics

	Recent CBCL sample (n=90)	Historical CBCL sample (n=98)	Recent YSR sample (n=84)	Historical YSR sample (n=123)
Sex				
Boys	45 (50%)	55 (56%)	44 (52%)	73 (59%)
Girls	45 (50%)	43 (44%)	40 (48%)	50 (41%)
Age at follow-up				
years ± sd	12.2 ± 1.5	12.7 ± 1.7	13.0 ± 1.7	14.4 ± 2.0
Cardiac diagnosis				
ASD	22	8	27	18
VSD	38	43	33	43
TGA	21	38	16	44
PS	9	9	8	18
Age at first invasive treatment				
ASD (years ± sd)	3.12 ± 1.32	2.28 ± 2.06	3.60 ± 1.42	3.88 ± 2.09
VSD	0.77 ± 1.10	0.63 ± 0.59	1.01 ± 1.60	1.32 ± 1.67
TGA	0.24 ± 0.60	0.58 ± 0.54	0.23 ± 0.66	0.74 ± 0.73
PS	1.28 ± 0.94	1.40 ± 1.40	1.79 ± 1.37	2.43 ± 1.87

Behavioural and emotional problems reported by parents

Analysis of variance for Total Problems, Internalising, Externalising, and the mean eight scale scores were performed in a group (recent sample versus historical sample) by sex (boys versus girls) by age (10-12 versus 13-15 years) factorial design. Table 2 shows the mean scale scores on the CBCL for both the recent and the historical sample for the eight scales, Internalising and Externalising and Total Problems. No significant differences were found between the mean problem scores reported by parents on the CBCL in the

recent versus the historical sample. No significant group by sex interactions or group by age interactions were found.

Next, ANOVAs were repeated, with cardiac diagnosis entered as a covariate. Cardiac diagnosis was adjusted for because the recent and historical CBCL sample differed significantly on cardiac diagnosis. The results did not change. These analyses of covariance did not reveal differences in parent-reports from the recent versus those of the historical sample.

Table 2. Mean problem scores on the CBCL parent-reports for the recent and historical patient sample

	Mean problem scores	
Scales	**Recent sample n=90**	**Historical sample n=98**
Anxious/Depressed	3.8	3.4
Withdrawn/Depressed	2.5	2.6
Somatic Complaints	2.2	2.2
Social Problems	3.7	3.7
Thought Problems	2.6	2.4
Attention Problems	3.7	4.4
Rule-Breaking Behaviour	1.4	1.6
Aggressive Behaviour	5.9	6.3
Internalising	8.5	8.3
Externalising	7.3	7.9
Total Problems	29.6	30.6

Self-reported behavioural and emotional problems

Table 3 shows the mean scale and mean Total Problems scores reported by patients in the recent and the historical ConHD sample, as well as the ANOVA results. On the YSR scale Withdrawn/Depressed, ConHD children in the recent sample reported significantly less problems than ConHD children in the historical sample. According to Cohen's criteria (Cohen, 1988) this group effect was small. A significant group x age interaction effect was found on the YSR for Somatic Complaints. This effect represented larger differences for the younger (11-13 years) versus the older (14-17 years) ConHD children in the historical sample compared to the recent sample, with the older ConHD patients scoring more favourably than the younger ConHD patients. No further significant two-way interactions were found.

Next, ANOVAs were repeated, now with cardiac diagnosis and age at follow-up entered as covariate in separate analysis. Again, these ANCOVAs did not show any further differences in self-reports from the recent versus the historical sample.

Table 3. Mean problem scores on the YSR for the recent and historical patient sample

	Mean problem scores	
Scales	**Recent sample n=84**	**Historical sample n=123**
Anxious/Depressed	3.9	4.1
Withdrawn/Depressed	2.4[a]	3.2[a]
Somatic Complaints	3.1[b]	2.9[b]
Social Problems	3.6	3.0
Thought Problems	3.3	2.9
Attention Problems	3.8	3.7
Rule-Breaking Behaviour	2.6	3.2
Aggressive Behaviour	5.3	6.3
Internalising	9.4	10.3
Externalising	7.9	9.5
Total Problems	32.4	33.6

[a] = 2.8 % of variance accounted for by significant group effect

[b] = 2.0 % of variance accounted for by significant group x age interaction

Discussion

The results of the present study showed that children with ConHD treated recently display the same level of behavioural and emotional problems compared to a sample of children with ConHD operated before 1980. The most prominent finding of this study is that despite evident improvements in diagnostic and surgical techniques and medical treatment of ConHD over the past decades, no clear improvement has been found in levels of behavioural and emotional problems as reported both by children who underwent invasive treatment for ConHD and their parents. No study thus far has compared long-term behavioural and emotional outcomes of a recent sample with those of a historical sample of children treated for ConHD by using the same standardized procedure, using well-known standardized parent- and self-reports.

Previous outcomes for the recent sample showed that according to parents' reports (CBCL) of problem behaviours, a significant proportion of ConHD children scored in the deviant range (16.9%) compared to the normative reference group (10.2%) (Spijkerboer et al., submitted b). According to patients themselves (YSR) the proportion of children and adolescents scoring in the deviant range, was comparable to that in the reference group (Spijkerboer et al., submitted b). For the historical sample, Utens et al. (1993) found that according to parent-reports the proportion of children and adolescents scoring in the deviant range was significantly greater in the ConHD sample (27%) compared to same-aged peers from reference groups (10%) at least 9 years after cardiac surgery.

At that time 20.7% of the YSR patient sample scored in the deviant range compared to 9.8% in the reference group (Utens et al., 1993). Remarkably, despite improvements in medical treatment for ConHD, parents of patients still evaluated their children's problem behaviours as significantly more unfavourable than the normative group. This conclusion can be drawn when looking at the proportion of patients from the historical and recent sample scoring in the deviant range.

The present study directly compared the level of behavioural and emotional problems from the recent and historical sample. From this approach we conclude that recent improvements in medical treatment are not reflected in more favourable behavioural and emotional outcomes for children treated for ConHD.

In the recent sample patients with an ASD, VSD and TGA were operated with standard cardiopulmonary bypass and patients with PS have undergone a balloon dilatation. In contrast, all patients in the historical sample underwent surgery. To what extent differences in operative procedures influenced our findings in unknown. The distribution of cardiac diagnoses and mean age at follow-up differed between the recent and the historical sample differed. When adjusted for these variables (as covariates), they did not appear to influence our results.

Based on the present study's findings we conclude that special attention should be given to screening and identifying children at risk for developing psychopathology, in order to provide them with adequate services and prevent the development of additional problems. Assessment and treatment of psychopathology at young age is desirable in ConHD patients, since psychopathological symptoms seem less persistent and thus better treatable at a young age.

Our results need confirmation in other medical settings, with other groups of patients with ConHD and with different cardiac diagnoses. Future research might also provide evidence for risk factors for the development of long-term behavioural and emotional problems in children with ConHD treated recently.

6

Medical predictors for long-term behavioural and emotional outcomes in children and adolescents after invasive treatment for congenital heart disease

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Abstract

Aims

To test the predictive value of medical variables for long-term behavioural and emotional problems in children and adolescents who underwent invasive treatment for congenital heart disease in infancy.

Methods

The Child Behavior Checklist (CBCL) was used to investigate to what extent parent-reported behavioural and emotional problems in 11-17-year-old congenital heart disease children can be predicted by: (1) medical history, (2) therapeutic intervention and direct post-interventional course, (3) long-term medical course, (4) present contact with physicians, and (5) present medical status.

Results

Higher CBCL Total Problems scores were predicted by cardiac medication prior to therapeutic intervention. Palliative intervention (Rashkind procedure) prior to therapeutic intervention was associated with more favourable scores on CBCL Total Problems and Externalising. The diagnostic categories atrial septal defect and pulmonary stenosis were associated with more favourable scores on CBCL Externalising compared to the group with a ventricular septal defect.

Conclusion

Long-term behavioural and emotional outcomes are only marginally predicted by medical variables. Cardiac medication prior to therapeutic intervention is a significant predictor for behavioural and emotional problems as reported by parents of patients. In counselling children with congenital heart disease and their parents, attention should be paid to children treated with cardiac medication prior to therapeutic intervention.

Introduction

Since mortality and morbidity in patients with congenital heart disease (ConHD) have improved significantly over the past decades, increasing attention is now being given to long-term behavioural and emotional outcome of these patients. It is important to know to what extent factors associated with the medical course in early childhood are predictive for long-term behavioural and emotional outcomes.

In the previous decades, several predictors for behavioural and emotional problems in children with ConHD have been identified, varying from maternal perceptions (DeMaso et al., 1991) to medical variables such as age at surgical repair, deep hypothermic circulatory arrest, and number of heart operations (Utens et al., 1998; Bellinger et al., 1997). Studies in which medical predictors of long-term behavioural and emotional problems are assessed in a recent sample of children treated for ConHD in the era after 1990, however, are lacking.

The present study is part of a follow-up study on long-term medical and psychosocial outcomes in children, adolescents and young adults who underwent invasive treatment for ConHD recently, that is between 1990-1995 (Spijkerboer et al., 2006; Spijkerboer et al., submitted b). In this follow-up we found that parents of ConHD children and adolescents evaluated the patients' behavioural and emotional problems, on average, as significantly more unfavourable than parents from a normative reference group (Spijkerboer et al., submitted b).

The aim of the present study was to determine which medical variables predicted long-term behavioural and emotional problems, in order to identify children and adolescents who are at risk for later maladjustment. We investigated to what extent long-term behavioural and emotional problems can be predicted by: (1) medical history, (2) therapeutic intervention and direct post-interventional course, (3) long-term medical course, (4) present contact with physicians, and (5) present medical status.

Methods

Outcome variables

The Child Behavior Checklist (CBCL) was used to obtain standardized parents' reports of behavioural and emotional problems in children aged 7-17 years (Achenbach, 1991a). The problem section consists of 120 problem items. Parents rate their child's behaviour during the preceding six months on a 3-point scale (0 = not true; 1 = somewhat or sometimes true; 2 = very true or often true). Good validity and reliability have been established (Achenbach, 1991a; Achenbach & Rescorla, 2001) and was confirmed for the Dutch version (Verhulst et al., 1996).

The CBCL consists of eight specific syndrome scales, and two broad band syndromes designated as Internalising and Externalising. Internalising problems reflect internal distress and externalising problems reflect conflicts with other people. The Internalising scale consists of the scales Withdrawn, Somatic Complaints and Anxious/Depressed, whereas the Externalising scale consists of the scales Rule-Breaking and Aggressive Behaviour. The syndrome scales Social Problems, Thought Problems, and Attention Problems belong neither to the Internalising nor the Externalising group. A Total Problems score can be obtained by summing the scores on all individual problem items. A higher score indicates a higher level of problems. For this particular study, the scales Internalising, Externalising and Total Problems were used.

Predictor variables

Table 1 shows five clusters of predictor variables, which were chosen on theoretical and/or medical grounds. Data were derived from medical examination during the follow-up and through retrospective medical file search. The variable hypothermia was dichotomised: 0 = moderate hypothermia (≥22°), 1 = deep hypothermia (<22°). The variable scar judged by physician was dichotomised: 0 = well healed, 1 = moderately or poorly healed. The variable sinus rhythm was dichotomised: 0 = sinus rhythm present, 1 = sinus rhythm absent. Most dichotomous prediction variables were coded as: 0 = no, favourable or risk absent and 1 = yes, unfavourable or risk present. The variable cardiovascular drug in the cluster present medical status was excluded since it occurred in only one patient. The variable restrictions imposed by physician was operationalized by one question for patients: Do you feel restricted by the physician? Patients treated with cardiac medication prior to therapeutic intervention received diuretics or prostaglandin E_1. Patients with a palliative intervention prior to therapeutic intervention underwent a Rashkind procedure.

Inclusion and exclusion criteria

During the follow-up, which took place in 2003-2004, consecutive surviving patients of 4 diagnostic groups, who underwent their first invasive treatment for ConHD between 1 January 1990 and 1 January 1996 in the Erasmus University Medical Centre Rotterdam, and who were younger than 15 years at the time of the treatment, were eligible. The sample in this study encompassed the following cardiac diagnostic groups: surgical closure of atrial septal defect (ASD), surgical closure of ventricular septal defect (VSD), arterial switch operation of transposition of the great arteries (TGA) and balloon dilatation for pulmonary stenosis (PS). Patients with proven syndromes of mental retardation, including Down syndrome, were excluded.

Patient sample

The target population consisted of 246 consecutive surviving patients. At follow-up 40 patients were lost (23 moved abroad, 17 were untraceable). Of the remaining 206 patients, 35 patients aged 18-28 years were not included since the CBCL did not cover this age-range. 171 patients were aged 7-17 years (age-range of the CBCL) at follow-up. From the CBCL sample parents of 46 patients refused to participate for practical or emotional grounds. Of these 46, 24 patients were aged 7-12 and 22 patients 13-17 years. Parents of 125 children aged 7-17 years completed the CBCL. One questionnaire was not usable because of incomplete information. The final CBCL sample consisted of parents of 66 male and 58 female patients (n=124). The response rate was 73%. Ten patients of this final CBCL sample did not participate in the medical part of the study. For a total of 64 boys and 50 girls (mean age 11.8 years, SD=2.4) both a CBCL parent-report and complete medical data were available. The number of patients in each diagnostic group was: surgical closure of ASD n=27 (mean age 13.9 years, SD=1.9), surgical closure of VSD n=43 (mean age 11.3 years, SD=2.2), arterial switch operation of TGA n=30 (mean age 10.4 years, SD=1.8) and balloon dilatation for PS n=13 (mean age 12.5 years, SD=2.7).

Assessment procedure

The research protocol was approved by the central committee on research involving human subjects before the start of the study. All patients were traced and approached uniformly. After an information letter was received, patients were called for an appointment by the research assistant. Before participating in the study, parents and/or patients signed an informed consent and returned it by mail. The definite cardiac diagnosis was checked by a paediatric cardiologist (W.H.). During the psychological investigation in the Erasmus University Medical Centre, parents of 7-17-year-old patients completed the CBCL in the waiting room whilst their child underwent a follow-up examination.

Statistical analyses

A three-phase strategy was followed, for each of the three outcome measures. The method of multiple linear regression analysis was applied. Since socio-economic status did not have a main effect on any of the outcome variables, it was not included in the models. In phase 1, each of the separate predictor variables was tested on the CBCL outcome (single 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 CBCL outcome (multiple analysis). The following clusters were used: medical history, therapeutic intervention and direct post-interventional course, long-term medical course, present contact 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 (for entry) and 0.25 (for removal). The procedure used was backward elimination procedure.

Table 1. Descriptive characteristics of the four diagnostic groups and the total patient sample on the predictor variables

Predictor variables	Cardiac diagnosis														
	ASD n=27			VSD n=43			TGA n=31			PS n=13			Total n=114		
	n	Mean	SD	n	Mean	SD	n	Mean	SD	n	Mean	SD	n	Mean	SD
Medical history															
Duration of pregnancy, in weeks		38.3	3.4		39.3	2.5		39.7	1.8		40.0	0.9		39.3	2.4
Weight at birth, in grams		2985.3	857.8		3056.5	721.7		3312.3	686.0		3444.5	411.5		3168.8	717.3
Cardiac medication prior to therapeutic intervention															
Yes	0			33			29			0			62		
No	27			10			2			13			52		
Palliative intervention prior to therapeutic intervention															
Yes	0			0			18			0			18		
No	27			43			13			13			96		
Therapeutic intervention and direct postinterventional course															
Age at first therapeutic intervention, in years		3.2	1.2		0.7	1.1		0.3	0.9		1.4	1.3		1.3	1.5
Hypothermia															
< 22°	1			1			7						9		
≥ 22°	26			42			24						92		
Direct postinterventional course (up to 2 weeks p.o.)															
With complications	2			1			4			0			7		
Without complications	25			42			27			13			107		
Time ICU, in days		1.0	0.4		1.9	1.6		4.8	5.1		0.1	0.3		2.4	3.4

Table 1, continued from previous page

Long-term medical course														
Number of hospitalisations as a result of heart problems since first therapeutic intervention														
≥ 1	0			3			1			1			5	
0	27			40			30			12			109	
Number of hospitalisations as a result of other problems														
≥ 1	13			18			15			4			50	
0	14			25			16			9			64	
Present contact with physicians														
Medical check-ups for the heart														
Once a year or more	0			3			1			1			5	
Less than once a year	27			40			30			12			109	
Restrictions imposed by physician														
Yes	1			3			2			1			7	
No	26			40			29			12			107	
Present medical status														
Scar judged by physician														
Moderately or poorly healed	3			2			5						10	
Well healed	24			41			26						91	
Maximum oxygen uptake		39.5	7.0		42.2	8.9		41.0	9.3		36.7	11.0		40.6	9.0
Sinus rhythm														
Absent	1			1			3			1			6	
Present	26			42			28			12			108	

Variables remaining in the regression model, applying to the clusters of predictor variables, were candidate-predictors for the final model. The final model in phase 3, p-values were set at 0.05 (for entry) and 0.051 (for removal), contained all significant variables from phase 2. Variables that showed significant results in this final model were considered the final predictors of CBCL outcomes. In order to correct for sex and age effects, sex and age were entered into each analysis in phases 1, 2 and 3. In order to prevent multicollinearity, the tolerance level (i.c. Variance Inflation Factor, VIF) was checked. The VIF was not allowed to exceed the value of 4. The linearity assumption was assessed by examining the scatter plots, with the continuous predictors on the x-axis and the dependent variables on the y-axis. The scatter plots showed that no other than linear relationships were to be expected. In the tables, the betas presented are standardized regression coefficients that express the relative importance of strength of the relationship between each predictor variable and the outcome variable. Plus versus minus sign indicates respectively the positive versus negative direction of the relation between the predictor variable and the outcome variable.

Results

Table 2 shows the results of phase 1 analyses. Cardiac medication prior to therapeutic intervention was significantly associated with a higher (that is an unfavourable) Externalising score, whereas the diagnostic categories of ASD and PS significantly predicted lower scores on Externalising. The variable scar judged by the physician as moderately or poorly healed was significantly associated with a higher Internalising score. Since phase 2 analyses served as a first selection of predictors and as an in-between model, the results are not presented here.

The results of the final model are presented in Table 3. Higher CBCL Total Problems scores were predicted by cardiac medication prior to therapeutic intervention. Palliative intervention prior to therapeutic intervention was associated with lower (that is favourable) scores on CBCL Total Problems and Externalising. The diagnostic categories of ASD and PS were associated with lower scores on CBCL Externalising.

Table 2. Prediction of outcomes on CBCL Total Problems, Internalising, Externalising by separate predictor variables adjusted for sex and age

Predictor variables	Total Problems	Internalising	Externalising
Medical history			
Duration of pregnancy, in weeks	-0.01	-0.01	0.04
Weight at birth, in grams	-0.08	-0.06	-0.09
Cardiac medication prior to therapeutic intervention	0.23	0.05	0.29*
Palliative intervention prior to therapeutic intervention	-0.22*	-0.18	-0.13
Cardiac diagnosis (reference: VSD)			
ASD	-0.19	<-0.01	-0.29*
TGA	-0.18	-0.00	-0.20
PS	-0.16	0.02	-0.23*
Therapeutic intervention and direct postinterventional course			
Age at first therapeutic intervention, in years	-0.19	-0.15	-1.32
Hypothermia	0.09	0.12	0.11
Direct postinterventional course	0.04	0.08	0.02
Time ICU	0.07	0.02	0.09
Long-term medical course			
Number of hospitalisations as a result of heart problems since first therapeutic intervention	0.11	<0.01	0.17
Number of hospitalisations as a result of other problems	0.08	0.10	0.03
Present contact with physicians			
Medical check-ups for the heart	-0.12	-0.09	-0.08
Restrictions imposed by physician	-0.08	-0.13	-0.01
Present medical status			
Scar judged by physician	0.11	0.20*	-0.01
Maximum oxygen uptake	-0.08	-0.23	-0.11
Sinus rhythm	-0.04	-0.06	0.02

* $P < 0.05$

Table 3. Final results of significant predictors of CBCL Total Problems and Externalising by main terms entered in the regression model

	Constant	Unstandardized coefficients β	95% CI	Standard error	Standardized coefficients β	*P*	Multiple R
Total Problems							
[a]Cardiac medication prior to therapeutic intervention	25.90	13.34	3.50 to 23.17	4.96	0.31	0.008	0.34
[a]Palliative intervention prior to therapeutic intervention		-16.49	-27.70 to -5.29	5.65	-0.28	0.004	
Externalising							
[a]Palliative intervention prior to therapeutic intervention	9.06	-3.80	-7.21 to -0.39	1.72	-0.22	0.029	0.32
[a]Cardiac diagnosis (reference category: VSD)							
ASD		-4.25	-7.65 to -0.84	1.72	-0.28	0.015	
PS		-4.46	-8.46 to -0.46	2.02	-0.22	0.029	

[a] Dichotomous predictor variables were coded as: 0 = no, favourable or risk absent, 1 = yes, unfavourable or risk present.

Discussion

The final prediction model of this study showed that cardiac medication prior to therapeutic intervention, was a significant predictor of long-term behavioural and emotional problems in 7-17-year-old children who underwent invasive treatment for congenital heart disease, as reported by their parents on the CBCL Total Problems score. Remarkably, palliative intervention prior to therapeutic intervention was a significant predictor for less long-term behavioural and emotional problems, as reported by parents on the Total Problems score and the Externalising scale.

Furthermore, the present study's results showed that patients who underwent a surgical closure of an ASD or a balloon dilatation for PS have a lower risk of developing externalising problems compared to patients who have been treated for a VSD. Apart for the externalising problems, the type of intervention (ASD, VSD and TGA treated by surgical intervention and PS treated by catheter intervention) is not significantly related to behavioural and emotional problems as reported by parents of the patients.

Several investigators have evaluated the effects of diagnosis. They did not find a relationship between the type and severity of the cardiac defect and the emotional (mal)adjustment of children with ConHD (DeMaso et al., 1990, 1991; Utens et al., 1998; Fredriksen et al., 2004). In a prediction study involving adults, van Rijen et al. (2004) also found that patients operated for a VSD have a higher risk of developing particularly externalising problems. We do not have a causal explanation for the higher risk of developing externalising problems in children with a VSD.

The only significant predictors found in this study all appeared to originate from the cluster medical history. Of the patients in our sample 54% (n=62) was treated with cardiac medication (diuretics or prostaglandin E_1) prior to therapeutic intervention. Table 1 showed that patients treated with cardiac medication were diagnosed with a VSD or TGA. Thus, cardiac medication prior to therapeutic intervention can be regarded as a marker for more serious cardiac illness. Eighteen patients with a TGA had a palliative intervention (Rashkind procedure) next to cardiac medication prior to therapeutic intervention.

It is difficult to explain why these medical variables separately had a different effect on long-term behavioural and emotional outcomes. The initial treatment of choice is related to the infants' underlying physical condition. From our data it can be assumed that children with more serious conditions received cardiac medication until surgical intervention could be performed. The time period that a child receives cardiac medication, in order to get well enough to undergo cardiac surgery, may be a period of fierce uncertainties and stress. This may influence parental perceptions regarding severity of the cardiac defect and coping of parents with the situation. Furthermore, it might negatively influence parent-child interactions. The time between diagnosis and therapeutic inter-

vention did not predict long-term behavioural and emotional outcomes. Why patients who were both treated initially with cardiac medication and a palliative intervention prior to the therapeutic intervention did not show more unfavourable long-term behavioural and emotional outcomes remains unclear. It is not known whether and to which extent side effects of cardiac medication can negatively influence later behavioural and emotional adjustment. As far as we know, this has not been reported. This topic should be investigated in future research.

In a prediction study executed about 15 years ago at our centre, Utens et al. (1998) found that the significant predictors identified at that time, such as a greater number of heart operations, deep hypothermic arrest, a short gestational age, and older age at surgical repair, also originated from the clusters of medical history and heart surgery. These significant predictors were not the same as those found in the present study. In the study population of Utens et al. (1998), however, all patients received surgical treatment. The TGA group was operated using the Mustard technique and PS patients were operated upon using inflow occlusion and valvotomy or with the use of cardiopulmonary bypass. Furthermore, patients with Tetralogy of Fallot and a miscellaneous diagnostic group were included. Predictors associated with higher Total Problems scores at that time, such as a greater number of heart operations and deep hypothermic arrest can be considered as relevant in their situation. In contrast with this, most of the patients in our sample were treated once (except of the group who had a palliative intervention prior to therapeutic intervention) and did not have reoperations.

A remarkable finding of our study is that variables concerning the present medical status and the present contact with physicians were not significantly associated with the occurrence of behavioural and emotional problems. Furthermore, we showed that behavioural and emotional problems are only marginally predicted by variables reflecting the medical history; these variables explain a maximum of 12% of the variance.

Previously, we found that according to parent-reports' (CBCL) of behavioural and emotional problems, a significant proportion of ConHD children scored in the deviant range (16.9%) compared to the normative reference group (10.2%) (Spijkerboer et al., submitted b). Therefore, in counselling ConHD children and their parents, it seems to be important to pay attention to patients who were treated with cardiac medication prior to therapeutic intervention. Assessment of behavioural and emotional outcomes in ConHD children may reveal problems that are not readily apparent in the routine clinical practice. It consequently may offer guidelines for counselling children with ConHD.

In conclusion our results indicate that, overall, the use of cardiac medication before surgical or interventional treatment was associated with unfavourable long-term behavioural and emotional outcomes. A palliative intervention prior to therapeutic intervention was associated with favourable long-term outcomes. Causal relationships need to be established.

7

Long-term psychological distress and coping styles in parents of children and adolescents who underwent invasive treatment for congenital heart disease

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Abstract

Aims

The level of psychological distress and styles of coping were assessed in both mothers and fathers of children and adolescents, who underwent invasive treatment for congenital heart disease (ConHD) at least 7 years and 6 months ago.

Methods

The General Health Questionnaire and the Utrecht Coping List were completed by parents of children with four different cardiac diagnoses.

Results

Overall, in comparison with reference groups, parents of children treated for ConHD showed lower levels of psychological distress, manifested as lower levels of somatic symptoms, anxiety and sleeplessness, and serious depression. Mothers of ConHD children reported significantly more somatic symptoms than fathers.

Further, compared to reference groups, ConHD parents showed more favourable outcomes; parents in our sample showed a weaker tendency to use coping styles such as reassuring thoughts and less often expressed negative emotions (anger, annoyance). Mothers of ConHD children appeared to seek social support more often compared to fathers.

Conclusion

Overall, lower levels of psychological distress and few differences in styles of coping compared to reference groups were found in parents of children treated for ConHD. We need to remain alert however, for individual parents at risk of adjusting poorly.

Introduction

As a result of important advances in diagnostic, surgical and catheter interventional techniques, the life expectancy of children with congenital heart disease (ConHD) has increased dramatically, and currently it is expected that more than 85% of infants born with ConHD reach adulthood (Bhat & Sahn, 2004).

The birth of an infant is an emotional moment for parents. Before and after the moment an infant is diagnosed with a ConHD parents experience a mixture of feelings. How parents respond to this situation, the diagnosis, hospitalisation, subsequent (surgical) intervention and caring for the infant at home, can affect both the short-term and long-term developmental outcomes of their children. Parents of newly diagnosed infants with ConHD have been reported to show heightened levels of stress compared with parents of healthy infants or parents of children newly diagnosed with cystic fibrosis (Goldberg et al., 1990b). Reactions of parents to an illness identified in infancy may be quite different from those occurring when children are older. Parental stress changes over time and is affected by the nature and course of the illness. Long-term coping styles may differ from those shortly after the disease was discovered. Nowadays, the long-term survival of children with ConHD has improved, but parents still have to cope with the effects of the illness on the functioning, development, and quality of life of their child, irrespective of the severity of the disease.

Many of the studies previously conducted on parents of children with ConHD focus on parental adjustment to the diagnosis and the short-term impact of the illness on the family, the early mother-infant relationship, and care-giving problems (Svavarsdottir & McCubbin, 1996; Goldberg et al., 1990a, 1991; Lobo, 1992). Thus, while there has been considerable attention to the short-term psychological effects of ConHD on parents, especially mothers, data on the long-term effect of ConHD on both parents are very limited.

The present study is part of a follow-up study concerning the long-term medical and psychological outcome in a cohort of children, adolescents and young adults who underwent invasive treatment for ConHD recently, that is between 1990 and 1995. The aims of the present study were:

1. To compare the levels of psychological distress and current coping styles of both mothers and fathers of 7-15-year-old children and adolescents who underwent invasive treatment for ConHD at young age, with that of reference groups.
2. To identify the role of sex, age and cardiac diagnosis in psychological distress and coping styles of parents of this sample.
3. To compare the level of psychological distress and coping styles of mothers versus fathers of the patient sample.

Methods

General Health Questionnaire

The 28-item version of the General Health Questionnaire was used. This is a reliable and valid standardized self-report, to assess the level of psychological distress (Koeter & Ormel, 1992). It contains four scales, which are concerned with: somatic symptoms, anxiety and sleeplessness, social dysfunctioning and serious depression. Cronbachs alpha for the total score is .94 (Koeter & Ormel, 1992).

Utrecht Coping List

We used the Utrecht Coping List, a reliable and standardized self-report to assess styles of coping (Schreurs et al., 1993). The respondent is asked to indicate how often he/she reacts to problems in a certain manner. 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 such as trying to relax by smoking or drinking), 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).

The satisfactory validity of the Utrecht Coping List has been described elsewhere (Schreurs et al., 1993). Construct validity and predictive validity has been examined by comparing the Utrecht Coping List with a range of other measures including those assessing coping, behaviour patterns, stress reactions, negative emotions, and personality variables.

Inclusion and exclusion criteria

During the follow-up, which took place in 2003-2004, consecutive surviving patients of 4 diagnostic groups, who underwent their first invasive treatment for ConHD at least 7 years and 6 months ago (between 1 January 1990 and 1 January 1996) in the Erasmus University Medical Centre Rotterdam, and who were younger than 15 years at the time of the treatment, were eligible. The sample in this study encompassed the following cardiac diagnostic groups: surgical closure of atrial septal defect (ASD), surgical closure of ventricular septal defect (VSD), arterial switch for transposition of the great arteries (TGA), and balloon dilatation for pulmonary stenosis (PS). Patients with proven syndromes of mental retardation including Down syndrome were excluded. Parents and patients had to be Dutch speaking. For the present study, parents of 7-15-year-old children were asked to participate.

Patient and parent sample

The target population consisted of 246 consecutive surviving patients. At follow-up 40 patients were lost (23 moved abroad, 17 were untraceable). Of the remaining 206 patients, parents of 159 patients in the age-range 7-15 years were eligible. Fifty sets of parents refused to participate on practical (n=39) or emotional grounds (n=11). For the remaining 109 patients, 52 sets of both parents, 48 mothers alone and 9 fathers alone, respectively, completed the General Health Questionnaire and 53 sets of both parents, and 50 mothers alone and 6 fathers alone, completed the Utrecht Coping List. The response rate was 69%.

Significant differences were found using Student's T-test between the mean ages of mothers (40.5 years, SD=4.7) and fathers (44.5 year, SD=5.7) of the General Health Questionnaire sample, and between the mean ages of mothers (40.5 years, SD=4.8) and fathers (44.6 year, SD=6.0) of the Utrecht Coping List sample.

Socio-economic status (SES) was scored on a 9-point scale of parental occupation, with scores 1 to 3 corresponding with elementary and so-called 'lower' occupations (SES-level 1), 4 and 5 corresponding with so-called 'middle' occupations (SES-level 2), and 6 to 9 corresponding with so- called 'higher' and scientific occupations (SES-level 3) (See Table 1) (Netherlands Central Bureau of Statistics, 1993).

The numbers of patients in each cardiac diagnostic group of the General Health Questionnaire sample was: surgical closure of ASD n=20 (mean age 13.2 years, SD=1.6); surgical closure of VSD n= 47 (mean age 11.3 years, SD=2.0); arterial switch operation of TGA n=31 (mean age 10.3 years, SD=1.6); and balloon dilatation for PS n=11 (mean age 11.9 years, SD=2.4). There were no differences as to the percentage of non-responders between the diagnostic groups.

Reference groups

Normative data were derived from the manuals of the questionnaires used. The reference groups for the General Health Questionnaire consisted of random samples drawn from the general population (Koeter & Ormel, 1992).

The male and female reference groups for the Utrecht Coping List consisted of employers of the Dutch railways and hospital nurses respectively, both combined with a random sample of the general population (Schreurs et al., 1993).

Assessment procedure

The research protocol was approved by the Dutch Central Committee on Medical Research involving Human Subjects before the start of the study. All parents and their children were traced, approached uniformly and signed an informed consent before participating in the study. The definite cardiac diagnosis was checked by a paediatric cardiologist (W.H.). During the psychological investigation of their child in the Erasmus

Table 1. Characteristics of patient and parent sample

	General Health Questionnaire n (%)	Utrecht Coping List n (%)
Age distribution of patients		
7-11 year	56 (51)	56 (51)
12-15 year	53 (49)	53 (49)
Gender distribution		
Boys	59 (54)	59 (54)
Girls	50 (46)	50 (46)
Cardiac diagnosis		
ASD, surgical closure	20 (18)	21 (19)
VSD, surgical closure	47 (43)	47 (43)
TGA, arterial switch	31 (29)	30 (28)
PS, balloon dilatation	11 (10)	11 (10)
Socio-economic status*		
SES-level 1	29 (27)	28 (26)
SES-level 2	51 (48)	53 (50)
SES-level 3	26 (25)	25 (24)
Total	109	109

Abbreviations. ASD: Atrial Septal Defect; VSD: Ventricular Septal Defect; TGA: Transposition of the Great Arteries; PS: Pulmonary Stenosis; SES: socio-economic status

* For 3 families SES data are lacking.

University Medical Centre, parents of 7-15-year-old children completed the General Health Questionnaire and Utrecht Coping List, independently from each other, in the waiting room. Most children were accompanied by one parent. If possible, the accompanying parent was asked to deliver the questionnaires to his/her partner, who could complete them at home and return them with a prepaid envelope.

Statistical analyses

All analyses were performed with two-sided tests; $p<0.05$ was considered significant. One sample t-tests were used to test differences in group means between the parents of the patient sample and reference groups on the General Health Questionnaire and Utrecht Coping List. To identify the role of sex (boys versus girls), age (7-11 versus 12-15 years) and cardiac diagnosis (ASD, VSD, TGA, PS) within the parent sample, univariate analyses of (co)variance, AN(C)OVAs were performed on all scales of the General Health Questionnaire and Utrecht Coping List. The variable socio-economic status (SES: 3 levels) showed a significant main effect in ANOVAs on all scales of the General

Health Questionnaire, indicating more psychological distress in the so-called 'lower SES classes'. Therefore it was applied as covariate in analysis on this questionnaire. On the Utrecht Coping List no main effects of SES were found. Differences in means on the General Health Questionnaire and Utrecht Coping List between mothers and fathers within families were assessed with paired t-test.

Results

Comparison of psychological distress of the parent sample versus reference groups

Table 2 shows the mean scores (and significant differences) on the General Health Questionnaire for the parents of the patient sample and reference group. Compared to reference females, mothers of children with ConHD reported significantly less complaints on the total score of the General Health Questionnaire, and the scales concerning somatic symptoms, anxiety and sleeplessness, and serious depression. Fathers of these children reported significantly less complaints than reference males with regard to the total score and all subscales, except social dysfunctioning.

Table 2. Mean scores on the General Health Questionnaire of parents of the patient sample and the reference group

	Parents of children with ConHD				Reference	
			Couples			
Scales	Mothers n=100	Fathers n=61	Mothers n=52*	Fathers n=52*	Females n=258	Males n=216
Somatic symptoms	4.6[1]	3.5[1]	4.7[1, 2]	3.7[1, 2]	6.7	5.4
Anxiety and sleeplessness	4.2[1]	3.9[1]	3.9[1]	4.1[1]	6.0	5.5
Social dysfunctioning	7.2	7.0	7.1	7.1	6.9	7.1
Serious depression	0.9[1]	0.5[1]	0.9[1]	0.5[1]	1.6	1.5
Total score	2.8[1]	1.8[1]	2.9[1]	2.2[1]	4.7	4.0

A high total score and high subscale scores is indicative of unfavourable functioning.

* For 52 parent couples, data were available from both parents.

[1] Significant (p<0.05) difference between mothers or fathers of children with ConHD versus same-sex reference groups.

[2] Significant (p<0.05) difference between mothers versus fathers within families.

Comparison of coping styles of the parent sample versus reference groups

Table 3 shows the mean scores on the Utrecht Coping List for the parents of the patient sample and reference group. Mothers of children with ConHD showed a weaker tendency to use palliative reaction, expression of negative emotions, and reassuring thoughts compared to the reference group. Compared to reference males, fathers of children with

ConHD showed a weaker tendency to use a passive reaction pattern, to express negative emotions, and to use reassuring thoughts.

Table 3. Mean scale scores on the Utrecht Coping List of parents of the patient sample and the reference group

	Parents of children with ConHD				Reference	
			Couples			
Scales	Mothers n=103	Fathers n=59	Mothers n=53*	Fathers n=53*	Females n=430	Males n=1048
Active problem solving	18.1	18.2	18.1	18.3	18.5	18.3
Palliative reaction	15.5[1]	15.2	15.7[1]	15.4	16.8	15.4
Avoiding / waiting	15.1	15.2	15.3	15.2	14.8	14.7
Seeking social support	13.4	11.2	13.5[2]	11.1[2]	13.9	11.2
Passive reaction pattern	10.2	9.8[1]	10.1	9.9	10.7	10.6
Expression of emotions	5.9[1]	5.8[1]	6.0[1]	5.8[1]	6.5	6.3
Reassuring thoughts	11.5[1]	10.6[1]	11.5	10.7[1]	12.1	11.5

On each scale, a high score indicates a strong tendency to use this coping style. A total score can not be computed.
* For 53 parent couples data were available from both parents.
[1] Significant (p<0.05) difference between mothers or fathers of children with ConHD versus same-sex reference groups.
[2] Significant (p<0.05) difference between mothers versus fathers within families.

Effect of sex, age and cardiac diagnosis within the parent sample

Parents of girls with ConHD reported significantly more complaints on the scale concerning serious depression of the General Health Questionnaire than parents of boys with ConHD. As to age and cardiac diagnosis no significant differences were found on any of the scales of the General Health Questionnaire within the parent sample. Neither any significant main effect was found for sex, age or diagnosis within the parent sample on any of the scales of the Utrecht Coping List. This means that on this list no difference was found between parents of male versus female patients, nor between parents of older versus younger patients, neither between parents of patients with ASD, VSD, TGA (surgical intervention) and PS (catheter intervention).

Comparison within families with children with ConHD: mothers versus fathers

On the General Health Questionnaire a significant difference within families with children with ConHD was found on the subscale somatic symptoms. Mothers reported significantly more somatic symptoms than fathers. On the Utrecht Coping List mothers scored significantly higher than fathers, concerning seeking social support. No further differences were found on either list.

Discussion

Level of psychological distress

Overall, the results of the present study showed lower levels of psychological distress in both mothers and fathers of children with ConHD compared to the reference groups.

In a previous study, using the same assessment instruments, Utens et al. (2002) found that parents of children who underwent elective cardiac surgery or cardiac catheter intervention also reported significantly lower levels of psychological distress than normative reference groups (Utens et al., 2002). In that study, the time of assessment of psychological distress in parents after the treatment of their children with ConHD differed from ours: parental reactions were assessed about 18 months after the cardiac procedure. In a more recent study Wray & Sensky (2004) found that both mothers and fathers of children with cardiac lesions had significantly higher rates of psychological distress prior to the surgical procedures than mothers or fathers of healthy children (Wray & Sensky, 2004). Twelve months following treatment, there was a significant reduction in the level of distress. Their sample was heterogeneous, with patients ranging in age from birth to 16 years, and covered a wide spectrum of cardiac diagnoses.

A possible explanation for our positive findings could be that the (acute), life-threatening nature of ConHD has decreased for the children and adolescents in our sample, since they underwent their first invasive treatment for ConHD at least 7 years and six months ago. A long-term stable medical status may relieve stress for many families. The frightening and stressful period, in which parents were confronted with the diagnosis and the cardiac procedure, is left behind them. Parents may have developed other reference norms and values than parents in the general population because of their past experience. The impact of this experience may have made parents stronger and could possibly lead to an attitude of worrying less about futilities in life.

To explain lower levels of psychological distress in parents after elective cardiac surgery or catheter intervention, Utens et al. (2002) referred to the possibility of the denial mechanism. In the study of Garson et al. (1978) denial was especially seen in parents of asymptomatic children (Garson et al., 1978). Denial may have influenced the present results too. Our response rate was satisfactory. However, to what extent data of non-responders would have led to less positive findings, is unknown.

Styles of coping

Overall, the styles of coping reported by parents of 7-15-year-old children with ConHD are comparable to those of same-aged reference adults in the general population. The significant differences found can be considered in favour of the parents in our sample: mothers and fathers of children with ConHD less often expressed negative emotions and used reassuring thoughts less often compared to the reference group. Further, mothers

of children with ConHD showed less palliative reactions and fathers of children with ConHD less often showed a passive pattern of reaction.

It can be speculated that past experiences (very difficult situations as cardiac surgery of one's child) may have led to different values and norms for parents of children with ConHD compared to the general population. This could possibly lead to less frequent use of reassuring thoughts by parents of patients. In contrast with our study, Wray & Sensky (2004) reported that mothers and fathers of children with cyanotic and acyanotic cardiac lesions used the coping mechanisms reassuring thoughts and active problem solving most frequently, and a mechanism of passive reaction least frequently. However, their methods differed from ours since they assessed parental coping styles more shortly after cardiac surgery of their child. Davis et al. (1998) found that maternal adjustment was associated with high levels of daily stress and palliative coping techniques (Davis et al., 1998). The study methods of Davis et al. (1998), however, differed from ours, since they used a different questionnaire to assess coping styles and the children of the mothers in their sample were two years or younger.

Effect of sex, age and cardiac diagnosis

Overall, in this study long-term psychological distress and styles of coping of parents of boys versus girls with different cardiac diagnosis (operated or treated by catheter intervention) are similar, apart from one exception. Parents of girls with ConHD reported more complaints on the scale concerning serious depression of the General Health Questionnaire than parents of boys with ConHD. As to age we found that parents of 7-11-year-old children with ConHD, showed no difference in psychological distress nor in styles of coping compared to parents of children aged 12-15 years.

Our results are favourable, considering the finding of Uzark & Jones (2003) that parenting stress tends to be higher in parents with older children. The mean age of the children with ConHD of the parents in their study was 6 years, ranging from 2 to 12 years. For parents it is more difficult to set limits for their child and maintain control with the child's advancing age. Past parental anxieties and overprotection with unnecessary restrictions can lead to future parenting problems, particularly during adolescence when children strive for independence (Uzark & Jones, 2003).

No differences in psychological distress nor in styles of coping between parents of the four diagnostic groups were found. This implicates that no differences were found between parents of patients who underwent cardiac surgery versus parents of patients who underwent catheter intervention (PS group). This is in line with previous findings, in which parental stress is unrelated to the severity of the child's heart disease (Uzark & Jones, 2003; Davis et al., 1998; Morelius et al., 2002). Results of the study of Wray & Sensky (2004) showed that, both before and after treatment, parents of children undergoing cardiac surgery or transplantation of bone marrow, as well as the parents of

healthy children, used the same mechanisms for coping, suggesting that neither the presence nor the nature of the chronic illness was influential in determining the type and frequency of strategies used.

In contrast with our findings, Utens et al. (2000a, 2002) reported that parents of children undergoing elective cardiac surgery showed more psychological distress, before and 18 months after the procedure, than did parents of those undergoing catheter intervention. However, results are difficult to compare, since different patient samples were used then, and the time of assessment differed. Our results might indicate that differences in stress between parents of surgically treated children and children treated by catheter intervention fade away on the long-term. However, since the number of patients who underwent catheter intervention was small, both in the present as well as in the previous studies, we should be careful in drawing firm conclusions.

Comparison within families with children with ConHD

Overall, no difference was found in psychological distress between mothers versus fathers of children with ConHD assessed at least seven years and six months after the first invasive treatment of their children, apart from one finding concerning somatic symptoms, mothers reported more distress than fathers.

Previous studies showed a higher level of distress in mothers of children with ConHD than fathers. Lawoko & Soares (2002) examined differences in distress (i.e. depression, anxiety, and somatisation) among parents of children with ConHD, parents of children with other diseases, and parents of healthy children (Lawoko & Soares, 2002). Mothers within all parent groups had higher levels of distress than fathers, with the highest level among mothers of children with ConHD compared to mothers in the other groups. Fathers of children with ConHD were doing worse than fathers belonging to the other groups. Utens et al. (2000a) found that mothers of children scheduled for elective surgery reported significantly more psychological distress than their husbands prior to the procedure. Goldberg et al. (1990b) compared responses of parents of infants with ConHD, those with cystic fibrosis, and those with healthy babies on the Parenting Stress Index. They reported that mothers reported more stress than fathers in most areas of the parent domain (these includes subscales for depression, attachment, parent health, role restrictions, sense of competence, relationship with spouse). In the general population a same trend was found: females showed more distress than men (Koeter & Ormel, 1992). In these previous studies, differences between mothers and fathers of children with ConHD were assessed more shortly after treatment than in our study.

Mothers of children with ConHD appeared to seek social support by sharing their feelings more often than fathers. This is in line with previous findings, in which mothers scored significantly higher with regard to seeking social support than fathers (Utens et al., 2000a). Wray & Sensky (2004) found in their study a significant decrease in the

use of social support for fathers of children with cyanotic cardiac lesions at follow-up compared with before surgery.

In our study socio-economic status showed a significant main effect, indicating more psychological distress in the so-called 'lower SES-classes'. The study of Uzark & Jones (2003) reported that parenting stress was not related to family socio-economic status.

Limitations of the present study

The present sample of children with ConHD contains four diagnostic groups, with specific age ranges, and is therefore not completely representative for all ConHD anomalies. Furthermore our sample of parents of children treated by catheter intervention was small, which restricts us in drawing firm conclusions comparing the treatment of children operated surgically or by catheter intervention. Though our response rate was satisfactory, psychological distress and coping styles could not be assessed in parents of all eligible patients. To what extent this may have influenced our data is unknown.

Implications

In an old, but classic qualitative study with 260 families, Garson et al. (1978) reported that the psychological needs of parents are not dependent on the seriousness of the heart disease of the child. They stated that children with less severe, i.e. mild congenital heart defects were at a higher risk for psychological problems because their parents frequently received less support from health providers than they needed. It has also been reported that overall improvements in the status of patients with ConHD may lead medical providers to underestimate the level of concern and stress experienced by their families (Van Horn et al., 2001). Parents need assistance to determine what is 'normal' for their child and how to monitor their child's health and safety (Sparacino et al., 1997).

A strength of the present study is that long-term psychological distress and coping styles were assessed in both mothers and fathers whose children underwent invasive treatment for ConHD. Overall, parents of the present patient sample showed favourable long-term outcomes. With progressing medical and technical possibilities, however, and more complex cardiac defects becoming operable, we need to remain alert for parents at risk of adjusting poorly.

Our sample was restricted to 7-15-year-old patients and older adolescents were not included. From literature it is known that the transition process to adult cardiology is a difficult phase (Sparacino, et al., 1997; Tong & Kools, 2004; Fernandes & Landzberg, 2004). As adolescents mature and display more independence, parents may struggle with letting go and leaving the responsibility to the adolescent. This process may lead to parental stress and adjustment of coping styles may be required.

It is known that the behavioural and emotional adjustment of children with ConHD is significantly related to the level of parenting distress (DeMaso et al., 1991). Gudmunds-

dottir et al. (1996) found that adolescents and young adults and their parents often used similar coping styles to gain control over the disease (Gudmundsdottir et al., 1996). Future research therefore should not only focus on short-term parental stress, but should also encompass long-term parental stress and coping styles, especially regarding the transition process. Insight in illness-related parental concerns creates the opportunity to improve clinical care. Awareness of the parental stress and coping styles enables health providers to support and facilitate the developmental and health transitions that both parents and their children experience.

8 General discussion

General discussion

So far, little is known about the long-term psychological outcomes in children with ConHD treated recently. In the present follow-up study several aspects of long-term psychosocial functioning were investigated in a sample of patients with four diagnostic groups, who underwent invasive treatment for ConHD between 1990 and 1995. Gaining insight in the long-term psychosocial outcomes is not only important for the diagnosed children themselves and their parents, but also for clinicians who work with these children. Assessment of psychosocial functioning may reveal important issues in the child or their parents that are not readily apparent in the routine clinical evaluations.

In this study several indicators of psychosocial functioning in children and adolescents with ConHD, such as health-related quality of life, intellectual functioning, and (school-related) behavioural and emotional outcomes were compared to the psychosocial functioning of children from normative reference groups. Psychosocial functioning of children with different cardiac diagnoses, both sexes and different ages was compared. A historical comparison was made between the level of long-term behavioural and emotional problems of patients treated recently and that of a historical sample of patients operated for ConHD in the same institute. Subsequently, the predictive value of medical variables, covering the complete medical course from birth up till now, for behavioural and emotional problems in the present patient sample, were examined. Finally, levels of psychological distress and current coping styles of parents of patients were compared with those of reference groups. In the current chapter the main findings and conclusions of the study will be discussed.

Present psychosocial functioning in children and adolescents with congenital heart disease

The total sample of ConHD children showed significantly lower HRQol scores regarding motor functioning, cognitive functioning, and positive emotional functioning compared to the reference group, reflecting poorer functioning in these domains for ConHD children.

Regarding intellectual functioning, the results showed that the mean WISC IQ- and factor scores were within the normal range, but the total sample of ConHD children obtained significantly lower scores on Verbal IQ and Verbal Comprehension compared to the reference group. In our sample 13% of the children were mentally retarded or had borderline intellectual functioning. The number of patients who had received special education services (14%) and had doubled in grade during lifetime (33%) indicates problems in a substantial portion of this population.

In previous studies, intellectual functioning was often assessed without measuring behavioural and emotional problems at school. Overall, in this study teachers' reports did not show elevated levels of school-related behavioural and emotional problems for ConHD children compared to reference children from the general population. ConHD children with poorer intellectual functioning showed more behavioural and emotional problems as reported by their teachers.

According to parents' reports on behavioural and emotional problems a significantly larger proportion of ConHD children obtained scores in the deviant range (16.9%) compared to the reference group (10.2%). According to patients themselves the proportion of children and adolescents scoring in the deviant range was comparable to that in the reference group. Parent-reports indicated significantly more total problems, more somatic complaints, more social, attention, and internalising problems for patients than for peers from the reference group. The relatively high level of somatic complaints reported for patients compared to the norm, could indicate that parents of ConHD children have an attitude to report these symptoms more often for their children due to alertness to illness related factors and bodily symptoms. It might also be explained by actual somatic problems in the ConHD patient sample. In contrast to the parent-reports, the self-reports of the 11-17-year-old ConHD patients showed favourable outcomes: reports were comparable to those of reference peers. Whether this is an underestimation of behavioural and emotional problems by patients themselves due to social desirability or denial, or a reflection of 'the truth' remains unclear.

A historical comparison was made between the level of behavioural and emotional problems of our sample of children and adolescents with ConHD treated recently, that is between 1990 and 1995, versus that of a comparable, historical sample operated between 1968 and 1980 in the same institute. The last decades many aspects of diagnostic, surgical and medical treatment for ConHD have been improved. The hypothesis was that these improvements would result in more favourable behavioural and emotional outcomes for children with ConHD treated recently. No previous study had ever made a historical comparison between the long-term behavioural and emotional outcomes of a recent sample versus those of a historical sample of children and adolescents treated for ConHD.

Results showed that children from the present sample, overall, displayed the same level of behavioural and emotional problems compared to children with ConHD operated before 1980. The most prominent finding was that despite evident improvements in medical treatment of ConHD, no clear improvement was found in levels of behavioural and emotional problems, as reported by both patients and their parents.

In conclusion, overall, our results showed that ConHD children showed a worse HRQol and a poorer intellectual functioning compared to reference groups. Elevated levels of behavioural and emotional problems were reported for ConHD children by their parents. Therefore, special attention should be given at screening and identifying children at risk.

Medical predictors

The medical variables which were significant predictors for long-term emotional and behavioural outcomes as reported by parents of 7-17-year-old patients originated from the cluster medical history. Cardiac medication (diuretics, prostaglandin E_1) prior to therapeutic intervention was associated with more unfavourable long-term behavioural and emotional outcomes. Palliative intervention (Rashkind procedure) prior to therapeutic intervention predicted more favourable long-term behavioural and emotional outcomes. Patients who underwent a surgical closure of an ASD or a balloon dilatation for PS had a lower risk of developing externalising problems compared to patients who had been treated for a VSD. We do not have a causal explanation for the higher risk of developing externalising problems in children with a VSD.

Of the patients in our sample 54% was treated with cardiac medication prior to therapeutic intervention. All these patients were diagnosed with a VSD or TGA. Cardiac medication prior to therapeutic intervention can be regarded as a marker for more serious illness. It is difficult to explain why cardiac medication and palliative intervention prior to therapeutic intervention had a different effect on long-term behavioural and emotional outcomes. The initial treatment of choice is related to the infants' underlying physical condition. The time period that a child receives cardiac medication, in order to get well enough to undergo cardiac surgery, may be a period of fierce uncertainties and stress. This may influence parental perceptions regarding severity of the cardiac defect and coping of parents with the situation. Further, it might negatively influence parent-child interactions. Whether and to which extent side effects of palliative medication can negatively influence later behavioural and emotional adjustment is not known. This should be investigated by future research. It can be concluded that since a maximum of only 12% of the variance was explained, behavioural and emotional problems are only marginally predicted by medical factors.

Psychological adjustment of parents

Overall, in comparison with reference groups, parents of patients showed lower levels of psychological distress, manifested as lower levels of somatic symptoms, anxiety and sleeplessness, and serious depression. No difference was found in psychological distress between mothers versus fathers of children with ConHD, apart from one exception: mothers reported more somatic symptoms than fathers. A possible explanation for our positive findings could be that the (acute), life-threatening nature of ConHD had decreased for the children and adolescents in our sample, since they underwent their first invasive treatment for ConHD at least 7 years and 6 months ago. A long-term stable

medical status may relieve stress for many families. The frightening and stressful period, in which parents were confronted with the diagnosis and the cardiac procedure, is left behind them. Parents may have developed other reference norms and values than parents in the general population because of their past experience. The impact of this experience may have made parents stronger and could possibly lead to an attitude of worrying less about futilities in life.

Overall, the styles of coping reported by parents of 7-15-year-old children treated for ConHD were comparable to those of same-aged reference adults in the general population. Furthermore, parents of patients expressed negative emotions (anger, annoyance) and used reassuring thoughts less often compared to the reference group.

It has been reported that the behavioural and emotional adjustment of children with ConHD is significantly related to the level of parental distress (DeMaso et al., 1991). Future research should therefore focus not only on short-term, but also on long-term parental stress and coping styles, especially regarding the transition process. From literature it is known that the transition to adult cardiology is a difficult phase (Sparacino, et al., 1997; Tong & Kools, 2004; Fernandes & Landzberg, 2004). Our sample was restricted to 7-15-year-old patients and older adolescents were not included.

The role of gender and age

ConHD children, aged 8-11 years, reported significantly poorer HRQol than reference peers on five domains: motor functioning, cognitive functioning, autonomy, social functioning and positive emotional functioning. Their parents reported significantly poorer HRQol regarding their children for cognitive functioning and positive emotional functioning compared to parents from the reference group. In contrast, reports of 12-15-year-old ConHD patients were, overall, comparable to those of reference peers (except on motor functioning), which is a favourable outcome.

Research into behavioural and emotional problems showed that, compared to a reference group, greater differences were found between younger (11-12 years) and older (13-17 years) children in the patient sample, with the younger patients scoring more unfavourably than the older patients.

Compared to reference children, ConHD children, aged 7-11 years, had significantly poorer intellectual functioning on four areas: Total IQ, Verbal IQ, Verbal Comprehension, and Perceptual Organization. Scores of 12-16-year-old ConHD children were, overall, similar to those of reference peers, which is a favourable outcome. Only one score, namely Verbal Comprehension appeared to be lower in this age group.

Patients, aged 8-11 years, showed a lower HRQol on positive emotional functioning than patients aged 12-15 years. Parents of patients aged 8-11 years reported poorer

quality of life regarding cognitive functioning than parents of ConHD children aged 12-15 years. Younger ConHD children, aged 7-11 years, showed a poorer intellectual functioning on Performance IQ and Perceptual Organization than ConHD children aged 12-16 years. This study showed that within the patient sample, the HRQol, the intellectual functioning and the level of behavioural and emotional problems of ConHD boys and girls were similar. Overall, our results showed that younger ConHD children had more unfavourable long-term psychosocial outcomes than older ConHD children.

The role of cardiac diagnosis

In this study, no significant differences were found between the HRQol and intellectual functioning of the different cardiac diagnostic groups. Neither any difference was found between the HRQol and intellectual functioning of patients treated by surgical intervention versus catheter intervention. In addition, our results showed no differences between the self-reports on emotional and behavioural problems of different cardiac diagnostic groups. According to parent-reports on behavioural and emotional problems, however, higher rates of social and externalising problems were found in patients treated for a VSD in comparison to other cardiac diagnostic groups

Research into prediction variables showed that patients with a VSD had a higher risk of developing externalising problems. Our results showed that the type of intervention is not significantly relevant in the prediction of behavioural and emotional problems, apart for externalising problems, as reported by parents of the present patient sample.

Overall, very few differences between cardiac diagnostic groups were found on the various indicators of psychosocial functioning. This is in line with previous findings (Fredriksen et al., 2004; Oates et al., 1995b; Utens et al., 1993).

Different informants

Overall, our results indicated that ConHD children reported poorer HRQol and reported more behavioural and emotional problems than their parents did about them. Our results regarding HRQol are in line with trends in the general population (Theunissen et al., 1998). Our results regarding the directions of discrepancies in child versus parent ratings of the level of behavioural and emotional problems resemble those of findings in the reference group. In the reference group, however, greater differences between child- and parent-reports were found. Considering our findings, we conclude that both ConHD patients themselves and their parents should be considered and used as important informants.

Strenghts and limitations

The present study encompasses survivors of four cardiac diagnostic groups treated between 1990 and 1995. Various aspects of psychosocial functioning were addressed, using structured and standardized assessment procedures. Also, medical predictors, covering the complete medical course from birth up till now, for long-term behavioural and emotional problems in the present patient sample were investigated. Furthermore, the hypothesis was tested that improvements in cardiological treatment would result in more favourable outcomes on behavioural and emotional problems for children with ConHD treated recently, in comparison to same-aged patients operated before 1980. In addition, psychological adjustment of parents was investigated. Overall, our results provided information regarding the long-term psychosocial functioning of different cardiac diagnostic groups of children and adolescents with ConHD that is not readily apparent from the routine clinical evaluations.

Besides these benefits, the study has some limitations. The present patient sample contained survivors of four frequently occurring diagnostic categories of patients. Patients with other diagnoses and patients with proven syndromes of mental retardation, including Down syndrome, were excluded. Therefore it may not be representative for all patients with ConHD. Patients were treated in one university hospital. To what extent results are generalizable to other centres is unknown.

Clinical implications

This thesis provides an overview on the long-term consequences for children after invasive treatment for congenital heart disease. This information can be used to guide adequate counselling and development of interventions aimed at enhancing patients' psychosocial functioning and their quality of life. The present study provides several clinical implications:

1. Results of this study show that the psychosocial functioning of children with ConHD is impaired to some extent. Therefore, screening of psychosocial functioning in children with ConHD next to the medical evaluation is recommended for clinical practice.
2. Assessment and treatment of psychosocial problems in patients with ConHD should be provided at an early age, since especially younger patients with ConHD are at risk for difficulties in psychosocial functioning.
3. In this study parent-child discrepancies were found on long-term HRQol and behavioural and emotional outcomes. We recommend use of multiple informants for valid and reliable assessment of HRQol and behavioural and emotional problems in children with ConHD.

Future research

This study revealed several outcomes on the long-term psychosocial functioning of children and adolescents after invasive treatment for ConHD, which might give direction to future studies.

Standardized, internationally well-known assessment procedures, such as used in this study should also be applied in other centers or in multi-center research. The present results need confirmation in other medical settings, with other groups of patients with ConHD and with different diagnoses. In addition, the effects of different therapeutic interventions (surgical treatment and catheter intervention) on the long-term psychosocial functioning should be studied in larger groups of patients. To investigate the stability of the present findings over time a longitudinal follow-up should be performed, for example, when all patients have reached adulthood.

Until now, little is known about side effects of cardiac medication prior to therapeutic intervention. The duration of the period of use could possibly negatively influence later behavioural and emotional adjustment. This deserves further exploration.

Furthermore, it is valuable to get more insight into the relation between impairments in physical functioning, especially motor functioning, and perceived HRQol.

In future, a disease-specific, cross-culturally widely applicable HRQol instrument should be applied in clinical practice of children with treated ConHD, in addition to generic quality of life measures, in order to better understand the impact of treated ConHD experienced by children and their parents and to be able to detect potential derangements in an early stage.

Future research should investigate the application and efficacy of evidence-based psychological interventions of psychosocial problems in children with ConHD.

To what extent the transition process to adult cardiology may influence parental adjustment should be further explored. Parents may struggle with letting go and leaving the responsibility to the adolescent. This process may lead to parental stress and adjustment of coping styles may be required.

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Summary

Summary

The objective of the study described in this thesis was to examine a range of long-term psychosocial outcomes of children and adolescents who underwent invasive treatment for congenital heart disease at a young age. In addition, the psychological adjustment of their parents was assessed.

In **chapter 1**, the background and the aims of this study were presented. Over the past decades, advances in diagnostic and surgical techniques and medical treatment of ConHD have gradually evolved and have significantly improved long-term survival of patients with ConHD. Despite improved survival, many patients with ConHD have more or less cardiac sequelae after surgical or interventional treatment. Next to survival, long-term morbidity is of great interest. Morbidity may impair the psychosocial functioning and quality of life of patients with ConHD. Hence, both clinicians and researchers have become more interested in the long-term psychosocial outcomes of patients treated for ConHD.

In the present study, various indicators of psychosocial functioning of children and adolescents with ConHD were measured. A follow-up examination was performed (from 2003-2004) among consecutive surviving patients of 4 diagnostic groups, who underwent their first invasive treatment for ConHD at least 7 years and 6 months ago (between 1 January 1990 and 1 January 1996) in the Erasmus University Medical Centre Rotterdam, and who were younger than 15 years at the time of the treatment. At follow-up 206 patients were aged 7-28 years. Of this group 35 patients were aged 18-28 years. Results of the 18-28-year-old patients were not included in this thesis. The present patient sample consisted of 171 patients, aged 7-17 years, and their parents.

The main aims of the present study were:

1. To compare the present psychosocial functioning of children and adolescents who underwent invasive treatment for ConHD at a young age with that of normative samples.
2. To determine the role of gender, age, and cardiac diagnosis on the psychosocial functioning of children and adolescents with ConHD.
3. To compare the level of long-term behavioural and emotional problems of the present sample of children and adolescents with ConHD treated recently, that is between 1990 and 1995, with that of a historical sample of children and adolescents operated for ConHD between 1968 and 1980 in the Erasmus University Medical Centre Rotterdam.
4. To determine the predictive value of a wide range of medical variables on long-term emotional and behavioural problems in children and adolescents with ConHD.
5. To compare the levels of psychological distress and current coping styles of both mothers and fathers of children and adolescents from the patient sample with that of reference groups.

In **chapter 2**, health-related quality of life was assessed with the TNO-AZL Child Quality of Life Questionnaire in both children and adolescents from our patient sample and their parents. The total sample of 8-15-year-old patients reported less favourable health-related quality of life regarding motor functioning, cognitive functioning, and positive emotional functioning compared to reference peers. Younger patients, aged 8-11 years, showed a worse quality of life regarding motor functioning, autonomy, cognitive functioning, social functioning, and positive emotional functioning than a same-aged reference group. Their parents reported poorer quality of life regarding them for cognitive functioning and positive emotional functioning compared to parents from the reference group. Overall, reports of 12-15-year-old patients were comparable to those of reference peers (except on motor functioning), which is a favourable outcome. The total sample of children with congenital heart disease reported significantly poorer quality of life than their parents did about them regarding pain and physical symptoms, motor functioning, autonomy and positive emotional functioning. No differences were found in quality of life between boys and girls with congenital heart disease, neither between different cardiac diagnostic groups. Overall, from our results it can be concluded that especially 8-11-year-old patients are at risk for poorer health-related quality of life, both according to their parents' as to their self-reports.

In **chapter 3**, long-term intellectual functioning and school-related behavioural outcomes of the present patient sample were examined by the Wechsler Intelligence Scale for Children-Revised and the Teacher's Report Form respectively. Overall, patients have IQ-scores that fall within the normal range. The total sample of 7-16-year-old children with congenital heart disease, however, obtained lower mean scores on Verbal IQ and Verbal Comprehension than reference children. Younger patients, aged 7-11 years, obtained significantly lower mean scores on Total IQ, Verbal IQ, Verbal Comprehension, and Perceptual Organization than same-aged reference peers. In contrast, scores of 12-16-year-old patients appeared to be significantly lower on Verbal Comprehension only and higher on Performance IQ compared to reference peers. No differences were found in intellectual functioning between boys and girls with congenital heart disease, neither between different cardiac diagnostic groups. In this study no differences were found between teachers' reports on school-related behavioural and emotional functioning of children with congenital heart disease versus those of reference children from the general population, apart from one exception. Overall, this sample of recently treated children with congenital heart disease and especially children aged 7-11 years, showed poorer intellectual functioning on several areas. These findings deserve further attention.

In **chapter 4**, the occurence of a wide range of behavioural and emotional problems in the patient sample were assessed. Patients (11-17-years-old) filled in the Youth Self-Report and parents of children with ConHD (aged 7-17 years) filled in the Child Behav-

ior Checklist. Parents of children and adolescents with ConHD evaluated the patients' behavioural problems as significantly more unfavourable than parents of the reference group. Parents reported significantly higher problems scores for the scales Somatic Complaints, Social Problems, Attention Problems, Internalising and Total Problems compared to parents from the reference group. In contrast, reports of patients were comparable to those of reference peers. No differences were found on the self-reports for different cardiac diagnostic groups. Discrepancies between self- and parent-reports were found, indicating that more problems were reported by ConHD patients themselves than by their parents.

Overall, parents of ConHD patients reported higher levels of behavioural and emotional problems compared to parents of the reference group, whereas patients themselves reported no long-term behavioural impairment. Younger ConHD patients deserve special attention. Assessing behavioural and emotional problems in ConHD patients can be helpful to detect children at risk for developing psychopathology.

In **chapter 5**, a historical comparison is made between the level of long-term behavioural and emotional problems in the present sample of children and adolescents with ConHD treated recently with that of a comparable, historical sample operated upon before 1980, also in the Erasmus University Medical Centre Rotterdam. The hypothesis was that improvements in cardiological treatment would result in more favourable outcomes on behavioural and emotional problems for children with ConHD treated recently, that is between 1990 and 1995, compared to same-aged patients operated before 1980. To assess behavioural and emotional problems of children and adolescents with ConHD the Child Behavior Checklist (parent-report) and the Youth Self-Report were used. The results show that parents and children from the recent ConHD sample reported fairly similar levels of behavioural and emotional problems compared to parents and children from the historical ConHD sample. Despite evident improvements in diagnostic and surgical techniques and medical treatment of ConHD over the past decades, virtually no changes have been found in levels of problem behaviour of children and adolescents who underwent invasive treatment for ConHD.

In **chapter 6**, the predictive value of medical variables, covering the complete medical course from birth up till now, for long-term behavioural and emotional outcomes in the present patient sample was examined. Behavioural and emotional problems were assessed by the Child Behavior Checklist, which was filled in by parents of patients. Higher CBCL Total Problems scores were predicted by cardiac medication (diuretics, prostaglandin E_1) prior to therapeutic intervention. Palliative intervention (Rashkind procedure) prior to therapeutic intervention was associated with lower (that is more favourable) scores on CBCL Total Problems and Externalising. Furthermore, the diagnostic categories atrial septal defect and pulmonary stenosis were associated with more favourable scores on CBCL Externalising compared to the group with a ventricular septal

defect. Long-term behavioural and emotional outcomes are only marginally predicted by medical variables. In counselling children with congenital heart disease and their parents, attention should be paid to children treated with cardiac medication prior to therapeutic intervention.

In **chapter 7**, the level of psychological distress and styles of coping in both mothers and fathers from the patient sample, were examined with respectively the General Health Questionnaire and the Utrecht Coping List. In comparison with reference groups, parents of children treated for congenital heart disease showed lower levels of psychological distress, manifested as lower levels of somatic symptoms, anxiety and sleeplessness, and serious depression. Mothers of congenital heart disease children reported more somatic symptoms than fathers. Parents in our sample showed more favourable coping styles compared to reference groups such as a weaker tendency to use reassuring thoughts and less frequent expression of negative emotions. Mothers of children with congenital heart disease appeared to seek social support more often compared to fathers.

Overall, lower levels of psychological distress and few differences in styles of coping compared to reference groups were found in the parents of children who underwent invasive treatment for congenital heart disease. We need to remain alert however, for individual parents at risk of adjusting poorly.

In **chapter 8**, the main findings and conclusions of this thesis were discussed. Overall, our results showed that ConHD children showed a worse HRQol and poorer intellectual functioning compared to reference groups. Parents of children with ConHD evaluated the patients' behavioural and emotional problems as significantly more unfavourable than parents of a reference group. Long-term behavioural and emotional outcomes are only marginally predicted by medical factors. Younger ConHD children showed more unfavourable long-term psychosocial outcomes than adolescents with ConHD. Very few differences between cardiac diagnostic groups were found on the various indicators of psychosocial functioning. Results showed that children from the present sample, overall, displayed the same level of behavioural and emotional problems compared to children with ConHD operated before 1980. Despite evident improvements in medical treatment of ConHD over the past decades, no clear improvement was found in levels of behavioural and emotional problems as reported by both patients and their parents. Overall, in comparison with reference groups, parents of patients showed lower levels of psychological distress. Few differences in styles of coping were found between parents of children treated for ConHD and reference groups. From the results of the present study it can be concluded that screening of psychosocial functioning in children with ConHD next to the medical evaluation can be recommended for clinical practice. Several clinical implications following from the results were described.

Samenvatting

Samenvatting

De hoofddoelstelling van het in dit proefschrift beschreven onderzoek was het bestuderen van een reeks psychosociale uitkomsten op lange termijn bij kinderen en adolescenten die op jonge leeftijd een invasieve behandeling voor een aangeboren hartafwijking ondergingen. Verder is de psychologische aanpassing van hun ouders onderzocht.

In **hoofdstuk 1** werden de achtergrond en de doelen van het huidige onderzoek geschetst. In de afgelopen decennia zijn diagnostische en chirurgische technieken en de medische behandeling van aangeboren hartafwijkingen geleidelijk steeds verder ontwikkeld. Dit heeft er toe geleid dat de lange termijn overleving van patiënten met een aangeboren hartafwijking enorm is verbeterd. Ondanks de sterk toegenomen overlevingsduur, hebben veel patiënten met een aangeboren hartafwijking nog in meer of mindere mate last van de cardiologische restverschijnselen na een chirurgische behandeling of interventie. Naast de overlevingsduur is de morbiditeit op lange termijn van groot belang. Morbiditeit kan het psychosociaal functioneren en de kwaliteit van leven van patiënten met een aangeboren hartafwijking aantasten. Hierdoor zijn zowel clinici als onderzoekers steeds meer geïnteresseerd geraakt in de lange termijn psychosociale uitkomsten bij patiënten die zijn behandeld voor een aangeboren hartafwijking. In het huidige onderzoek werden verschillende indicatoren van psychosociaal functioneren bij kinderen en adolescenten met een aangeboren hartafwijking gemeten. In 2003 en 2004 werd een follow-up onderzoek uitgevoerd bij de overlevende patiënten van 4 diagnostische groepen, die tenminste 7 jaar en 6 maanden geleden (tussen 1 januari 1990 en 1 januari 1996) in het Erasmus MC te Rotterdam hun eerste invasieve behandeling voor een aangeboren hartafwijking hadden ondergaan en die ten tijde van de behandeling niet ouder waren dan 15 jaar. Tijdens de follow-up waren 206 patiënten tussen de 7 en de 28 jaar oud. Onder hen bevonden zich 35 patiënten tussen de 18 en 28 jaar. Resultaten van de 18 tot 28-jarige patiënten werden niet in dit proefschrift opgenomen. De huidige onderzoeksgroep bestaat uit 171 patiënten tussen de 7 en 17 jaar en hun ouders.

De doelstellingen van dit onderzoek waren:

1. Het vergelijken van het huidige psychosociaal functioneren van kinderen en adolescenten die op jonge leeftijd een invasieve behandeling ondergingen voor een aangeboren hartafwijking met dat van normgroepen.
2. Het bepalen van de rol van geslacht, leeftijd en cardiale diagnose ten aanzien van het psychosociaal functioneren bij kinderen en adolescenten met een aangeboren hartafwijking.
3. Het vergelijken van het niveau van gedrags- en emotionele problemen op lange termijn in de huidige groep kinderen en adolescenten die recentelijk, dat is tussen 1990 en 1995, zijn behandeld voor een aangeboren hartafwijking met dat van een

historische groep kinderen en adolescenten die tussen 1968 en 1980 eveneens in het Erasmus MC te Rotterdam zijn geopereerd voor een aangeboren hartafwijking.
4. Het bepalen van de voorspellende waarde van een groot aantal medische variabelen op emotionele en gedragsproblemen op lange termijn bij kinderen en adolescenten in de huidige onderzoeksgroep.
5. Het vergelijken van het niveau van psychologisch onwelbevinden en de huidige coping stijlen bij zowel moeders als vaders van kinderen en adolescenten van de huidige onderzoeksgroep met dat van normgroepen.

In **hoofdstuk 2**, werd de gezondheidsgerelateerde kwaliteit van leven van de patiëntengroep gemeten door afname van de TNO-AZL Kwaliteit van Leven Vragenlijst bij zowel kinderen en adolescenten als hun ouders. De totale groep van 8-15 jarige patiënten rapporteerde een slechtere gezondheidsgerelateerde kwaliteit van leven voor motorisch functioneren, cognitief functioneren en positief emotioneel functioneren vergeleken met leeftijdgenoten uit de normgroep. Jongere patiënten, tussen de 8 en 11 jaar, lieten een slechtere kwaliteit van leven zien voor motorisch functioneren, autonomie, cognitief functioneren, sociaal functioneren en positief emotioneel functioneren dan leeftijdgenoten van de normgroep. Hun ouders rapporteerden een slechtere kwaliteit van leven over hen wat betreft cognitief functioneren en positief emotioneel functioneren vergeleken met ouders uit de normgroep. De rapportages van 12-15 jarige patiënten waren vergelijkbaar met die van leeftijdgenoten uit de normgroep (behalve voor wat betreft motorisch functioneren); dit is een gunstige uitkomst. De totale huidige patiëntengroep rapporteerde een significant slechtere kwaliteit van leven dan hun ouders deden over hen voor wat betreft pijn en lichamelijke symptomen, motorisch functioneren, autonomie en positief emotioneel functioneren. Er werden geen verschillen gevonden in kwaliteit van leven tussen jongens en meisjes met een aangeboren hartafwijking en evenmin tussen verschillende cardiale diagnostische groepen. Over het geheel genomen, kan worden geconcludeerd dat 8-11 jarige patiënten, zowel volgens de ouder- als de zelfrapportages, een verhoogd risico lopen op een slechtere kwaliteit van leven.

In **hoofdstuk 3** werden het intellectueel functioneren en de schoolgerelateerde gedragsuitkomsten op de lange termijn onderzocht bij de huidige onderzoeksgroep met respectievelijk de herziene versie van de Wechsler Intelligentie Test voor Kinderen (WISC-R) en de Teacher Report Form. Over het geheel genomen behaalden de patiënten IQ-scores die binnen de normale range lagen. De totale groep van 7-16 jarige kinderen met een aangeboren hartafwijking behaalde echter lagere gemiddelde scores voor het verbale IQ en verbaal begrip dan kinderen uit de normgroep. Jongere patiënten, tussen de 7 en 11 jaar, behaalden significant lagere gemiddelde scores voor totaal IQ, verbaal IQ, verbaal begrip en perceptuele organisatie dan een even oude normgroep. In tegenstelling hiermee, bleken scores van 12-16 jarige patiënten alleen significant lager

voor verbaal begrip en hoger voor performaal IQ, vergeleken met leeftijdsgenoten uit de normgroep. Er werden geen verschillen gevonden in intellectueel functioneren tussen jongens en meisjes met een aangeboren hartafwijking, noch tussen verschillende cardiale diagnostische groepen. In dit onderzoek werden geen verschillen gevonden in rapportages van leerkrachten wat betreft schoolgerelateerd gedrags- en emotioneel functioneren van kinderen met een aangeboren hartafwijking in vergelijking met de rapportages voor kinderen uit de algemene bevolking, op één uitzondering na. Over het geheel genomen, liet onze onderzoeksgroep op verschillende gebieden een slechter intellectueel functioneren zien. Deze bevindingen verdienen verdere aandacht.

In **hoofdstuk 4** werd het voorkomen van een groot aantal gedrags- en emotionele problemen op lange termijn gemeten in de huidige onderzoeksgroep. Patiënten (tussen 11-17 jaar) vulden zelf de Youth Self-Report in en ouders van patiënten (tussen de 7-17 jaar) vulden de Child Behavior Checklist in over hun kinderen. Ouders van kinderen en adolescenten van onze onderzoeksgroep beoordeelden de gedragsproblemen van hun kinderen als significant ongunstiger dan ouders in de normgroep. Ouders van de huidige onderzoeksgroep behaalden significant hogere probleemscores op somatische klachten, sociale problemen, aandachtsproblemen, internaliserende- en totale probleemscores in vergelijking met de normgroep. De scores van patiënten zelf waren echter vergelijkbaar met die van leeftijdgenoten uit de normgroep. Er werden geen verschillen gevonden tussen de probleemscores van verschillende cardiaal diagnostische groepen. Verschillen tussen de zelf- en ouderrapportages gaven aan dat patiënten met een aangeboren hartafwijking meer problemen rapporteerden dan hun ouders deden over hen. Over het algemeen rapporteerden ouders van de patiëntengroep meer gedrags- en emotionele problemen in vergelijking met de normgroep, terwijl de patiënten zelf op lange termijn geen verschillen met de normgroep lieten zien wat betreft probleemgedrag. Jongere patiënten met een aangeboren hartafwijking verdienen extra aandacht. Het onderzoeken van gedrags- en emotionele problemen bij patiënten met een aangeboren hartafwijking is nuttig voor het opsporen van kinderen die een verhoogd risico lopen op het ontwikkelen van psychopathologie.

In **hoofdstuk 5** werd een historische vergelijking gemaakt tussen het niveau van lange-termijn gedrags- en emotionele problemen van de huidige onderzoeksgroep die recentelijk is behandeld voor een aangeboren hartafwijking en dat van een vergelijkbare historische onderzoeksgroep die voor 1980 ook in het Erasmus MC te Rotterdam is geopereerd voor een aangeboren hartafwijking. De hypothese luidde dat verbeteringen in cardiologische behandeling zouden resulteren in meer gunstige uitkomsten wat betreft gedrags- en emotionele problemen bij kinderen die recentelijk, dat is tussen 1990 en 1995, zijn behandeld voor een aangeboren hartafwijking, vergeleken met even oude patiënten die voor 1980 zijn geopereerd. Gedrags- en emotionele problemen werden gemeten met de Child Behavior Checklist (ouder-rapportage) en de Youth Self-Report.

De resultaten laten zien dat ouders en kinderen in de huidige onderzoeksgroep een vrijwel gelijk niveau van gedrags- en emotionele problemen rapporteerden in vergelijking tot ouders en kinderen in de historische onderzoeksgroep. Ondanks duidelijke verbeteringen in diagnostische en chirurgische technieken en medische behandeling van aangeboren hartafwijkingen in de afgelopen decennia, worden er vrijwel geen veranderingen gevonden in het niveau van probleemgedrag bij kinderen en adolescenten die een invasieve behandeling voor een aangeboren hartafwijking hebben ondergaan.

In **hoofdstuk 6** werd de voorspellende waarde van medische variabelen, die het hele medische beloop van de geboorte tot nu bestrijken, voor de huidige lange-termijn gedrags- en emotionele uitkomsten in de patiëntengroep onderzocht. Gedrags- en emotionele problemen werden gemeten met de Child Behavior Checklist, die werd ingevuld door de ouders. Hogere CBCL Totale Probleemscores werden voorspeld door cardiale medicatie (diuretica, prostaglandine E_1) gebruikt gedurende de periode voorafgaand aan de therapeutische interventie. Het ondergaan van een palliatieve interventie (Rashkind procedure) voorafgaand aan de therapeutische ingreep bleek gerelateerd aan lagere, dat wil zeggen gunstigere uitkomsten, op CBCL Totale Probleemscores en Externaliseren. Verder bleken de diagnoses atrium septum defect en pulmonaal stenose geassocieerd met gunstigere scores op CBCL Externaliseren in vergelijking tot de groep met een ventriculair septum defect. Gedrags- en emotionele uitkomsten op lange termijn werden slechts marginaal voorspeld door medische variabelen. In de medische behandeling van kinderen met een aangeboren hartafwijking en hun ouders dient aandacht geschonken te worden aan kinderen die zijn behandeld met cardiale medicatie voorafgaand aan de therapeutische interventie.

In **hoofdstuk 7** werden het niveau van psychologisch onwelbevinden en de coping stijlen van zowel moeders als vaders van kinderen en adolescenten in onze patiëntengroep onderzocht door middel van respectievelijk de General Health Questionnaire en de Utrechtse Coping Lijst. In vergelijking met normgroepen, vertoonden ouders van kinderen die zijn behandeld voor hun aangeboren hartafwijking een lager niveau van psychologisch onwelbevinden, hetgeen weerspiegeld wordt in minder somatische symptomen, angst, slapeloosheid en ernstige depressie. Moeders van kinderen met een aangeboren hartafwijking rapporteerden meer somatische symptomen dan vaders. Ouders van kinderen met een aangeboren hartafwijking vertoonden gunstigere coping stijlen in vergelijking met normgroepen zoals het minder vaak gebruiken van geruststellende gedachten en het minder vaak uiten van negatieve emoties. Moeders van kinderen met aangeboren hartafwijkingen leken in vergelijking met vaders vaker sociale steun te zoeken. Over het geheel genomen, werden lagere niveaus van psychologisch onwelbevinden en weinig verschillen in coping stijlen gevonden tussen ouders van kinderen uit onze patiëntengroep in vergelijking met normgroepen. We dienen echter

alert te blijven op individuele ouders die een risico lopen op een slechte psychologische aanpassing.

In **hoofdstuk 8** werden de belangrijkste bevindingen en conclusies van dit proefschrift besproken. Over het geheel genomen, lieten de kinderen met een aangeboren hartafwijking een slechtere gezondheidsgerelateerde kwaliteit van leven en een slechter intellectueel functioneren zien vergeleken met normgroepen. De ouders van onze onderzoeksgroep beoordeelden de gedrags- en emotionele problemen van hun kinderen als significant ongunstiger dan ouders in een normgroep. Gedrags- en emotionele uitkomsten op lange termijn werden slechts marginaal voorspeld door medische variabelen. Jongere kinderen met een aangeboren hartafwijking hadden minder gunstige psychosociale uitkomsten op lange termijn dan adolescenten in de onderzoeksgroep. Er werden weinig verschillen gevonden op de verschillende indicatoren van psychosociaal functioneren tussen de verschillende cardiale diagnostische groepen. Patiënten in de huidige onderzoeksgroep lieten over het geheel genomen een gelijk niveau van gedrags- en emotionele problemen zien in vergelijking tot kinderen met een aangeboren hartafwijking die voor 1980 waren geopereerd. Ondanks forse verbeteringen in de medische behandeling van aangeboren hartafwijkingen in de afgelopen decennia, werd er geen duidelijke verbetering gevonden in het niveau van gedrags- en emotionele problemen zoals gerapporteerd door patiënten en hun ouders. Over het geheel genomen, vertoonden ouders van kinderen die waren behandeld voor hun aangeboren hartafwijking een lager niveau van psychologisch onwelbevinden in vergelijking met normgroepen. De coping stijlen van ouders van de patiëntengroep lieten weinig verschillen zien met die van referentiegroepen. Op grond van de huidige onderzoeksresultaten kan geconcludeerd worden dat screening van psychosociaal functioneren bij kinderen met een aangeboren hartafwijking aan te bevelen is naast de cardiologische controle voor de klinische praktijk. Verschillende uit de resultaten voortkomende klinische implicaties worden beschreven.

Dankwoord
Curriculum Vitae

Dankwoord

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♥ Alma

Curriculum Vitae

Alinda Wilma Spijkerboer (roepnaam: Alma) werd geboren op 18 mei 1975 te Putten. In 1993 behaalde zij haar VWO diploma aan de Pieter Zandt scholengemeenschap te Kampen. In hetzelfde jaar begon zij met de studie Sociale Wetenschappen aan de Universiteit Utrecht, waar het doctoraal psychologie, in de richting ontwikkelingspsychologie werd behaald in juli 1998.

Vanaf januari 1999 tot en met maart 2000 werkte zij op de afdeling Kinder- en Jeugdpsychiatrie van het UMC Utrecht mee aan een onderzoek naar de genetische achtergronden van kinderen met ADHD en aanverwante gedragsstoornissen.

Vanaf april 2000 tot en met januari 2003 was zij als psycholoog-onderzoeker werkzaam binnen de divisie Jeugd & Ouderen van Altrecht (instelling voor geestelijke gezondheidszorg), locatie Zeist. Tevens volgde zij in 2002 de opleiding Moderne Bedrijfsadministratie.

Vanaf februari 2003 tot maart 2006 was zij als research-psycholoog aangesteld op de afdeling Kindercardiologie (hoofd: prof.dr. W.A. Helbing) en de afdeling Kinder- en Jeugdpsychiatrie (hoofd: prof.dr. F.C. Verhulst) van het Sophia Kinderziekenhuis - Erasmus MC te Rotterdam. In deze periode werd een onderzoek uitgevoerd naar psychosociale uitkomsten op lange termijn bij patiënten die op jonge leeftijd een invasieve behandeling hebben ondergaan voor een aangeboren hartafwijking (projectleiders: prof.dr. W.A. Helbing en mw.dr. E.M.W.J. Utens), waarvan de resultaten in dit proefschrift beschreven zijn.