

The Psychosocial Consequences of a Congenital Heart Disease in Adult Life: Young Female Patients at Risk

Elisabeth M.W.J. Utens,¹⁾ Elisabeth H.M. van Rijen,²⁾
Jolien W. Roos-Hesselink,³⁾ Folkert J. Meijboom,⁴⁾ and Frank C. Verhulst¹⁾

¹⁾Department of Child and Adolescent Psychiatry,
Erasmus MC - Sophia Children's Hospital, ²⁾Institute of Psychology,
Department of Social Sciences, Erasmus University Rotterdam,
³⁾Department of Cardiology, Thoraxcenter, Erasmus MC, Rotterdam,
⁴⁾Department of Congenital Cardiology, University Medical Centre,
St. Radboud, Nijmegen, The Netherlands

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This review describes the main findings of a long-term longitudinal cohort study investigating the psychosocial functioning of adults with congenital heart disease (ConHD). This longitudinal study encompasses two follow-ups of (almost) the same cohort of patients.

Methods: For this study, all consecutive patients who underwent their first open heart surgery for ConHD between 1968-1980 in the Erasmus Medical Center Rotterdam, and who were younger than 15 years of age at the time of surgery, were eligible. The first follow-up was executed between 1989-1991 and the second follow-up, of the same cohort, between 2000-2001 (follow-up intervals respectively: 9-23 years and 20-33 years after the first cardiac surgery).

Results: Overall, in both follow-ups, favourable outcomes were found as to biographical variables and social functioning. No clear relationships were found between cardiac diagnoses and psychosocial outcomes. In the second follow-up, significantly more ConHD patients scored in the psychopathological range compared to the normative reference group. Especially young female patients (20-27 years) were at risk for elevated levels of psychopathology. Young female patients showed less adequate coping styles and they felt more limited due to physical functioning than older female patients.

Conclusion and Discussion: Young female patients may have struggled with disease specific uncertainties as to sexual relationships, birth control, pregnancy risks, delivery and offspring, which may have negatively influenced their emotional functioning.

Clinical Implications Encompass: early screening and treatment for psychopathology, attention to subjective restrictions, patient education, monitored physical training and psychological counselling by a psychologist with expertise in this very unique field.

Introduction

With the enormous advances of medical treatment of congenital heart disease (ConHD), increasing survival rates of patients with ConHD have resulted in a new and fastly growing patient population: adults with operated ConHD. These patients not only experience the consequences of their heart condition, but also of having grown up with a congenital anomaly. Since knowledge about specific psychosocial

problems and needs of adults with ConHD was limited, in Rotterdam two long-term follow-up studies were executed in (almost) the same cohort of patients to investigate the quality of life and psychosocial functioning in these adult patients.

This article aims to provide a short review of the main findings of the long-term "Rotterdam follow-up studies into Quality of Life" of patients operated for congenital heart defects in early childhood. Result will be presented as to:

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Reprint requests to: E.M.W.J. Utens, PhD., Department of Child and Adolescent Psychiatry,
Erasmus MC - Sophia Children's Hospital
Dr. Molewaterplein 60, PO Box 2060, 3000 CB Rotterdam, The Netherlands

- 1) psychosocial outcomes in adults with congenital heart disease;
- 2) psychopathology in (young female) adults with ConHD;
- 3) finally, in the conclusion/discussion the focus is on disease specific topics (such as scars, sexual functioning and pregnancy) and clinical implications for treatment and counselling adults with ConHD.

This review is based on the lecture given by Elisabeth Utens at the 44th Annual Meeting of the Japanese Society of Pediatric Cardiology and Cardiac Surgery (For detailed description of the different study results: see references).¹⁻⁸⁾

Rotterdam Follow-up Studies into Quality of Life

1. Psychosocial Outcomes in Adults with ConHD

1. First and second Rotterdam follow-up study: overall outcome

In the longitudinal Rotterdam cohort study, all consecutive patients who underwent their first open heart surgery for ConHD between 1968-1980 in the Erasmus Medical Center Rotterdam, and who were younger than 15 years of age at the time of surgery, were eligible. The medical and psychological follow-up examinations were done in hospital, by a congenital cardiologist and a psychologist.

The first follow-up^{1, 2)} was executed between 1989-1991. The second follow-up, of (almost) the same cohort, took place between 2000-2001.³⁻⁷⁾ Follow-up intervals were respectively: 9-23 years and 20-33 years, after the first cardiac surgery. Four hundred and ninety-eight patients participated in the first study (response rate: 87%). Three hundred and sixty-two of these patients participated in the second follow-up. At the second follow-up, the age-range was: 20-46 years (response rate: 90%).

Main conclusion: overall, in both follow-up studies, favourable psychosocial outcomes were found (as to biographical variables and social functioning) and remarkably no clear relationships were found between cardiac diagnoses and psychosocial outcomes.

2. Second Rotterdam follow-up study (2000-2001): psychosocial outcomes³⁻⁷⁾

Since the second follow-up is the most recent study, results of only this follow-up will be described below. In this second follow-up a miscellaneous diagnostic group (N = 61) consisting of (very) small numbers of patients with a variety

of congenital heart defects was now excluded. Included were again patients of the five larger diagnostic groups were: atrial septal defect (N = 93), ventricular septal defect (N = 97), tetralogy of Fallot (N = 77), transposition of the great arteries (N = 55), and pulmonary stenosis (N = 40). A miscellaneous diagnostic group (N = 61) consisting of (very) small numbers of patients with a variety of congenital heart defects was now excluded.

Favourable outcomes were found on the variables: living conditions, marital status, offspring rate and social functioning. "Favourable" here indicates that the patient sample showed high similarity with reference groups on these variables (It should be noticed that statistical analyses were done for different age categories and gender separately).³⁾

Living conditions: The living conditions of the patient sample were comparable to those of normative reference groups. 78% was living independently, that is not with parents. *Marital status:* Marital status was favourable, that is: comparable to normative data: 72% had a stable relationship.

Offspring: The offspring rate was assessed for both married/cohabitant patients, both for men and women. None of the 20-24 years old patients had any children, compared to 14% in the normative reference group. Patients between 25-39 years of age showed normal offspring rates. Thus patients seemed to raise families somewhat later in their twenties compared to a reference group. For patients older than 40 years of age, the offspring rates were lower compared to those for younger patients; so after 40 years offspring rate dropped.

Social functioning: Outcomes as to social functioning, that is leisure time activities, were comparable to or even better (more favourable) than in reference groups.

Unfavourable outcomes were found as to educational and occupational levels.³⁾ The proportion of adult patients with a history of special education was high: 27%. Of these, 85% had attended schools for learning-disabled or mentally handicapped, 15% had attended schools for chronically ill children. Accordingly patients showed lower educational and occupational levels compared to reference groups. 85% had a paid job, 7% received a social security benefit and 8% still attended school or took full-time care of the household.

Results as to cardiac diagnoses.³⁾ Overall, no significant differences were found between cardiac diagnostic groups as to: living conditions, marital status, offspring rate, educational attainments, occupational levels and receiving social security benefits. Patients with: transposition of the great ar-

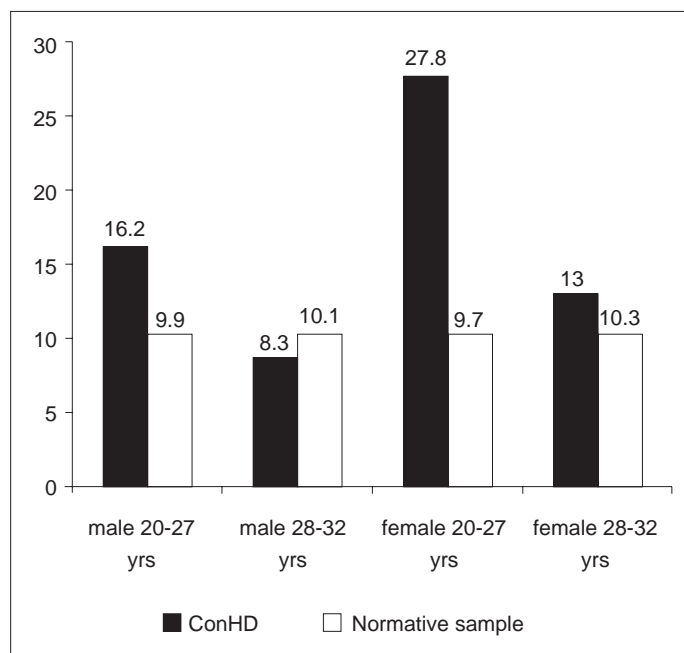


Fig. 1 Percentages in psychopathological range.

teries and tetralogy of Fallot, however, showed some sequelae reflected by weaker poorer performance on education, occupation and intensive sports, but further were able to lead normal lives.

2. Psychopathology in (Young Female) ConHD Adults

1. Second Rotterdam follow-up study (2000-2001): psychopathology⁴⁾

1) Methods

Psychopathology was assessed with two internationally well-known questionnaires, developed by Achenbach⁹⁾: the Dutch version of the Young Adult Self-Report (YASR), to be completed by patients themselves, and the Young Adult Behaviour Checklist (YABCL), to be completed by a significant other (parent or spouse) regarding the patient. Both lists contain (± 110) parallel problem items. A total problem score, an internalizing and externalizing score can be computed. Internalizing consists of the scales: anxiety/depression, withdrawal and somatic complaints. Externalizing consists of: intrusive, aggressive and delinquent behaviors. In addition, thought and attention problems are assessed. A high score is unfavourable and indicate the presence of problems.

2) Patients

The YASR was completed by 349 patients (188 males, mean age: 30 years, range: 20-46 years, response rate: 88%). The YABCL was filled in by 334 parents and/or spouses (response rate: 84%).

3) Results

Percentages in psychopathological range

Both patients' self-reports (YASR) and significant others' reports (YABCL) showed significantly higher percentages of patients who scored in the deviant, psychopathological range compared to the normative reference group. Parents and partners of patients rated the patients' behaviour as even more deviant from the norm than patients themselves, on a broad range of problem areas.

Unfavourable outcomes for female patients: psychopathology⁴⁾

Fig. 1 shows results on the YASR; normative data were available for the age-range 20-32 years. Especially young female patients (20-27 years) showed higher levels of psychopathology (27.8%) compared to the same-aged reference females (9.7%). These younger female patients also showed higher levels of psychopathology compared to older female patients (28-32 years). Differences between younger and older women were larger in the patient sample than in the reference group. Finally, female patients showed more psy-

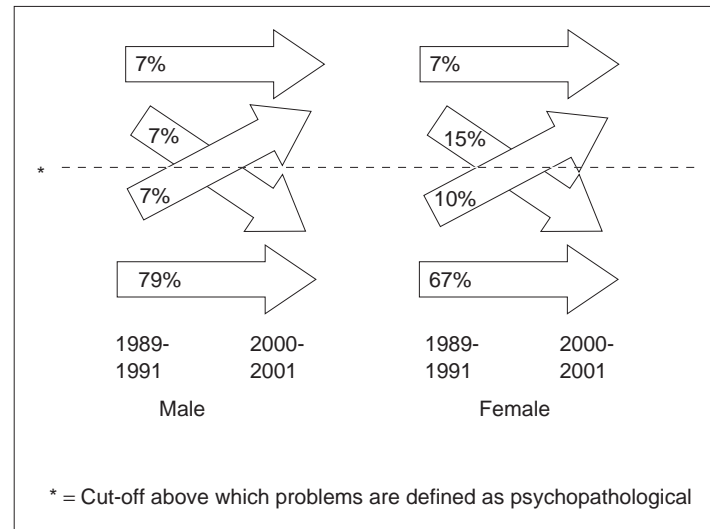


Fig. 2 Trajectories in psychopathology of ConHD adults over a 10 year period.

chopathology than male patients; these effects of gender were also larger in the patient sample than in the reference groups.

Trajectories in psychopathology of ConHD adults over a 10 year period⁴⁾

In the first and second follow-up study the same questionnaires were used, thus providing the possibility to assess longitudinal trajectories in psychopathology over a 10 year period. In Fig. 2 the dotted line indicates the cut-off above which problems are defined as psychopathological. The arrows indicate the percentages of patients, who in comparison to 10 years ago, have persisted in their level of psychopathology and scored above the cut-off at both follow-ups; patients for whom problems have decreased respectively have increased and patients who remained stable at a so-called normal level (below cut-off). Fig. 2 shows that female and younger patients showed more variability in trajectories of psychopathology than did male and older patients. Overall behavioural and emotional problems diminished when the female patients become older.

Predictors of psychopathology⁵⁾

In the second follow-up, significant predictors for the elevated level of psychopathology reported by patients were: being female, restrictions by the scar as felt by patients, low exercise capacity and physical restrictions imposed by physicians. Regarding the scar, subjective experiences of patients appeared to be a better predictor for late problems

than objective judgments by physicians. When significant others (parents) were the informants: early hospitalisations with re-operations were predictive for long-term problems.

Further unfavourable outcomes for female patients: coping and limitations^{3,6)}

Finally, in the second follow-up it was found that, compared to reference women, ConHD female patients showed less favourable coping styles (for example less active problem solving). Younger female patients reported more role limitations due to physical functioning than older female patients. As to feelings about having offspring, 44% of the female patients reported that their heart condition somehow had been a limiting factor in the choice of having children (For the male patients this percentage was 20%).⁸⁾

Conclusion and Discussion

In conclusion, the adults patients from this second follow-up study overall seemed capable of leading normal lives and motivated to make good use of their abilities (apart from some residual effects for patients with tetralogy of Fallot and transposition of the great arteries). Especially young female patients were at risk for psychopathology and less adequate coping. They experienced more behavioural- and emotional problems and felt more limited due to physical functioning than older female patients.

An explanation for the present findings may be that young female patients struggle with aspects of life, such as

establishing and maintaining a stable sexual relationship and having offspring, whereas these aspects seem less prominent later in adulthood. For young female patients of the present follow-up the surgical scars may have resulted in feelings of uncertainty, unattractiveness and fear of rejection, when entering intimate relationships.

With regard to surgical scars a Canadian study from Kañtoch et al.¹⁰⁾ showed that 60 of their 100 patients felt permanently disfigured by the scar and (nearly) half of them concealed it. A median sternotomy was not perceived as more disfiguring than lateral thoracotomy or submammary incision. Multiple scars, wide scars and those with keloid formation or chest deformity were considered to be more disfiguring. Scars were reported to have a negative impact on self-confidence in 23% of women and 13% of men. Chest scarring, however, had a limited effect on sexual relationships. Moreover, scars had a positive impact on appreciation of health in 60% of the patients, since the scars reminded patients them to value their health.

For female patients from the present study disease specific uncertainties as to sexual relationships, birth control, pregnancy risks, delivery and offspring may have negatively influenced their emotional functioning. All these issues involve their own bodily functioning and may cause anxiety about their physical functioning. The more passive coping styles of young female patients found in the present study, may reflect feelings of having less control and greater vulnerability as to the obstacles and barriers in life they encounter. Many of our female patients felt restricted by the ConHD in their choice of having children. This may explain why our patient sample seemed to raise families later than the general population.

These disease-specific uncertainties described above may also explain why our younger female patients showed more limitations as to physical health compared to our older female patients. Older female patients may have already overcome these uncertainties once they have established a stable relationship and/or a satisfactory family life.

Clinical implications

Considering the findings described above, the following guidelines for counselling adults with congenital heart disease can be given.

• Screening and treatment for psychopathology

Screening and treatment for psychopathology in adult patients with ConHD, especially in young female patients,

should be provided at an early stage, since at young age these symptoms are less persistent and better treatable. With psychological treatment of these problems and underlying disease specific uncertainties, the struggle into adulthood may become less hard and thus the patients' quality of life may be improved.

• Attention to subjective restrictions

In assessing the patients' well-being, special attention should be given to restrictions young ConHD female patients experience as to the scar, sexual functioning, fertility status, birth control, pregnancy risks, heritability, raising children and life expectancy. Subjective restrictions due to the scar should be evaluated instead of the more objective, esthetical aspects. In Belgium, Moons et al.¹¹⁾ found that adult ConHD patients reported fewer sexual problems than control persons. In his study however, ConHD women experienced distress at "being insecure about having sex, not enjoying sex and not being aroused" more often than male patients. Clinical experience learns that fear of death due to arousal and overexertion may exist. Inability to have children due to cardiac risks may be a devastating blow to a woman's sense of identity and self-worth (Horner et al.).¹²⁾ All these issues may cause behavioural and emotional problems.

• Disease-specific patient education

Disease-specific patient education on the above topics may relieve uncertainties, misconceptions and unnecessary burdens or limitations. Anxieties should be directly addressed. Due to fear of sudden death, patients may avoid long-term relationships and future planning. A review from Hargrove et al.¹³⁾ shows that many young people have poor understanding of their heart condition, especially of sexual and reproductive health issues, whereas they are just as likely as healthy peers to be sexually active (Horner et al.).¹²⁾ In Germany respectively in the United Kingdom Vigl et al.¹⁴⁾ and Rogers et al.¹⁵⁾ found that many ConHD female patients received poor or no advice about contraception or pregnancy risks from their treating physician. Correct information should be given to avoid risks of unplanned pregnancy. In Canada Reid et al.¹⁶⁾ found that 48% of 212 young ConHD adults (19-29 years old) was sexually active (that is: had at least one partner in the previous 3 months). This rate was lower compared to that in normative samples. Of these adult patients 36% engaged in potentially risky sexual behaviours (having had multiple partners, questionable birth control, using drugs or alcohol before sex). Women with complex ConHD had the most concerns about fertility, heredity,

pregnancy risks. This indicates the need for disease-specific patient education.

• **Monitored physical training**

Monitored physical training might help patients with poor physical condition to feel more secure about their bodily function.

• **Psychological counselling**

Cardiologists should have the opportunity to refer patients with psychosocial problems to psychologists who are specialized in this very unique field. Psychological counselling can enhance active coping styles in female patients so that they can feel more in control of their situation. This is a worthwhile aim to strive for.

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