A Life Less Ordinary
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Chapter 10
General discussion and future perspectives
The studies described in this thesis were undertaken to increase our insight in how CHD influences socio-economic aspects in adult life and how this can be improved.

EMPLOYMENT AND EDUCATION

Employment in adults with congenital heart disease (CHD) was investigated in several ways. With the help of in-depth interviews and different questionnaires, we identified different occupational challenges faced by adults with CHD in daily life. Patterns in job participation for men and women were assessed in adults with CHD and the general population in a sub study of the study of Zomer et al. Furthermore, we studied different aspects of employment including work ability and limitations at work in the Assessment of Patterns of Patient-Reported Outcomes in Adults with Congenital Heart disease (APPROACH) – International Study (IS) and the COgnition in COngenital heart disease (COCO) study.

Although the majority of adults with CHD is working, employment rates often differ from the general population and are generally lower. This appears to be influenced by disease-specific aspects such as CHD severity and comorbidity, patient-specific aspects such as performance status, educational level, sex or age and country-specific aspects such as health care and social security structure. Limitations at work are often reported. Although risk factors for unemployment and limitations at work are often similar to those in other chronic diseases, certain CHD-specific limitations are also mentioned. Examples are progressive symptoms in extreme weather, telling (or not telling) the employer about the CHD and hiding (or not hiding) visual signs of the CHD.

In the Netherlands, currently around 70% of adults with CHD is employed. This is lower than in the general population. We found employment rates varying from 66% in our random sample from CONCOR for the insurance study to 71% among Dutch participants of APPROACH-IS (N=254). Five years before, Zomer et al. found an employment rate of 66% in adults with CHD (N=1496). Compared to the general Dutch population, all the employment rates we found for adults with CHD in the Netherlands are reduced, although the difference varies in magnitude. In an effort to exclude selection bias from different reference groups, we also compared the employment rates in adult CHD patients to data from Eurostat, the statistical office of the European Union. Eurostat reports employment rates of 77% in 2010 and 75% in 2014 in the age group 20–64 year.

Only a few countries have reported similar or even higher levels of employment among CHD patients. What could be the recipe to their success? A Finnish study found comparable educational levels and employment levels even higher than expected among adult CHD survivors. In this study, only patients with cyanotic defects were less likely to obtain a university degree. As an explanation for the high employment rate, the authors refer
to the high value CHD patients attributed to work as described by Horner et al. from qualitative data. It remains unclear why these findings among American patients would only apply to these specific patients. Ternestedt et al. followed patients with tetralogy of Fallot (TOF) and atrial septal defect (ASD) up to 30 years after surgery and found educational levels higher than the Swedish general population. At follow-up, almost everyone was successfully employed. The authors explain their positive results by referring to CHD patients as ‘high achievers’. Although not confirmed by all results from the neuropsychological examination in our COCO study, some of our results (on the Stroop and TMT-A test) supported this ‘high achievers’ pattern. However, this coping strategy is undoubtedly not used by all CHD patients.

Caruana et al. found similar education and employment rates in ACHD patients compared to the general population in Malta. They also found a significantly higher number of male CHD patients with higher-skilled jobs than in the general population. The explanation proposed by the authors is that Maltese men are generally more often involved in lower-skilled jobs, and that CHD patients have probably been encouraged to pursue a less physical job, which often needs higher education. In both our qualitative study as the study from Sparacino et al., patients repeatedly addressed the subject of how their CHD had influenced their parent’s educational advice. Some parents supported a higher education because of the physically less strenuous future jobs, other parents emphasized less on good education because of the CHD and sometimes uncertain future.

One could state that our COCO study, a study on cerebral damage and employment, would attract patients with working problems. However, even in the APPROACH-IS, where patients did not expect questions on work, the same high proportion of patients with limitations at work was found. In both studies, with a majority of employed patients, limitations at work were reported by approximately one third of patients. In our qualitative study, limitations were mentioned by almost all patients. These limitations were not without consequences. Half of the patients who experienced limitations had been absent from work in the prior 3 months because of work-related problems. Even among patients without reported limitations, several patients reported absence from work during the prior 3 months due to their cardiac condition.

APPROACH-IS also showed that unemployment was associated with lower QOL. Differences in QOL (measured by the LAS, a linear analogue scale ranging from 0 to 10) between employed and unemployed patients per country are shown in Figure 1. Only in India and Taiwan, no difference in QOL was seen among employed or unemployed patients. The largest effect from employment on QOL was seen in Belgium and Sweden.
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**Figure 1.** Mean difference in QOL (expressed by LAS) between employed and unemployed CHD patients

Abbreviations: QOL = quality of life; LAS = linear analogue scale; CHD = congenital heart disease

**PRICES TO PAY FOR IMPROVED SURVIVAL**

**Cerebral damage**

Cerebral damage could be a possible explanation for the unemployment and limitations at work observed in adult CHD. Patients with CHD are at increased risk for developmental disorders. This could be caused by several factors, associated with either the CHD or treatment itself or more environmental circumstances. Especially cyanotic defects and defects requiring multiple surgeries, seem to be at risk for cerebral damage. Although cerebral damage has frequently been reported in new-borns and children, little about this is known in adults. Furthermore, cerebral damage is often measured by structural imaging as MRI or by cognitive assessment (NPE). We studied cerebral damage through both measurements in adults with corrected tetralogy of Fallot (TOF), the most common cyanotic CHD. Although a large part of patients showed signs of cerebral damage and a large part of patients experienced limitations at work, there were no connections between the three, as shown in Figure 2.
Cerebral infarcts were seen in 12 of 64 patients (19%). These were predominantly silent infarcts, considering only 1 of those patients had a neurological history. Besides this, white matter hyperintensities (WMH) were seen in 29 patients (45%). The clinical relevance of the findings we call ‘damage’, is not clear. In fact, we show with the lack of a neurological history and no relation with neuropsychological or occupational outcomes at all, that at this time, damage seems limited to scientific observations. Our major limitation was the lack of an age-related reference group. To our knowledge, the prevalence of WMH in this age group is unknown, especially when measured on a sensitive 3-Tesla MRI scanner. However, according to several clinical experts, WMH and infarcts are generally not seen in this age group.

Cerebral damage was related to age and age of corrective surgery. Patients with cerebral damage were significantly older at the time of their first surgery. More important, for each year older when considered surgically corrected, 14% more cerebral damage was seen. Nowadays, primary repair of TOF at young age (between 6 and 18 months) is common practice. This study supports studies that are in favour of early surgery regarding cerebral outcome. Although we found no indication that this cerebral damage had any clinical or occupational relevance, with the current younger ages of corrective surgery, we expect less cerebral damage. Furthermore, higher educational levels are also described among TOF patients that have been operated at lower ages.
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Cancer risk

Clinical observations from daily practice suggest an increased cancer risk in CHD, partly supported by three other studies in this field 14-16. We did not find an increased cancer incidence in adults with CHD. In the other studies on cancer and CHD, only one study found an increased cancer incidence, but this was mainly in children with CHD 14. The increased cancer incidence in the other study was driven by Down syndrome and disappeared when they were excluded 15. Another recent study reported an increased cancer prevalence in Canada compared to the general population 16. Our results suggest that the earlier reported increased incidence could be mainly a problem among children with CHD, that cancer risk factors are much more favourable in adults with CHD or that we have underestimated cancer incidence in our cohort due to left censoring.

Favourably, most studies show healthier lifestyles of CHD patients compared to controls, most notably less smoking in adults with CHD, which causes a substantial part of cancers 1, 17, 18. Unfavourably, most patients with CHD receive considerable doses of ionizing radiation 19-23. It is often received during childhood, the most sensitive time to harmful effects and often at sensitive localisations at the body 20, 23-27. Although radiation doses for specific procedures have decreased over the years, the use and frequency of examinations using radiation is still increasing as well as the amount of interventional therapeutic catheter procedures, the latter often resulting in even more radiation exposure. However, to show an effect on cancer risk, patients (including their risk factors) have to be followed for long periods of time. Also, large groups are needed since cancer incidence at these young ages is low. Given the different methods and low cancer incidence in these age groups, we already did an effort to combine data from CONCOR and other CHD cohorts in an international study. Unfortunately, because of the different ways the CHD cohorts were composed and cancer data were acquired, so far this has not led to a uniform database. We have learned that ideally, future research of cancer incidence in CHD should include data on lifestyle and other cancer risk factors, preferably including information on radiation exposure. With the recent launch of KinCOR, the Dutch national registry for children with CHD, these limitations will hopefully be overcome in the future 28.

Quality of Life (QOL)

An important subject in ‘prices to pay for improved survival’ is QOL. APPROACH-IS showed that worldwide, QOL in adults with CHD is generally good. There was no reference group, but when results for the Netherlands were compared with general population data from Statistics Netherlands (CBS), QOL appeared to be even higher in adults with CHD (LAS 8.0 in adults with CHD versus 7.8 for the general population) 29. The average score for QOL in Europe was 7.1.
FUTURE PLANS TO IMPROVE EDUCATION AND EMPLOYMENT IN ADULT CHD

What could lower the risk of unemployment and work limitations? The observations of lower employment rates and frequent limitations at work, encouraged to think of possible solutions. Advice on education and career should be given more often. Crossland et al. showed it improves employability. However, advice should be useful and therefore be given by someone experienced in CHD as well as job coaching.

Although cardiac rehabilitation programs are well known and established in other cardiac diseases, this type of interventional programs in adult CHD patients are still not implemented in daily practise. A specific program that focuses on improving occupational outcomes is, as far we know, lacking at all. However, the integrated multidisciplinary approach of a cardiac rehabilitation program might be highly suitable.

In other chronic somatic diseases, self-management programmes led by a professional trainer and a trained patient suggest to promote self-care, improve patients’ self-efficacy, self-reported health, cognitive symptom management and frequency of exercise. Still there is no evidence that such self-management programs could improve occupational outcomes. However, in this specific group of adults with CHD who experience limitations at work, a targeted self-management program might help CHD patients to overcome the barriers or limitations reported. The CHAPTER 2 study is a Canadian example of a tailored self-management program, which tried to minimize the transition time from paediatric to adult care for CHD patients by using pre-defined goals, led by a trained nurse. A similar design may also be used to address certain well-described problems in adult CHD patients regarding education and employment. Our results emphasize that such programs should focus on dealing with symptoms, effective communication with colleagues and employer and gaining insight in internal and external recovery options.

Finding and maintaining a good balance in work and personal life is a challenge for most people these days, especially when you need more time to recover from work.

Another tool could be the so-called ‘personalised care planning’. This is a process used in the management of several (non-CHD so far) chronic diseases in which patients and clinicians collaborate to identify problems caused by or related to the patient’s condition, in order to develop a plan to improve these problems. A specialised adult CHD centre will presumably have the best structure to facilitate such planning.

The work ability score (WAS) was used as a tool to determine work ability in our study. In other chronic diseases, it has proven a useful tool to predict future disability. Especially for patients that experience limitations at work but are still working, it could help identify patients who are about to be absent from work to intervene, in order to avoid absenteeism. The question is who should do this. In several of our studies, we noticed that it is unclear who feels responsible for preventing absenteeism. Besides this, patients often feel that telling the employer about their CHD might be contra productive and most
patients only see a company doctor or occupational physician when absence is already an issue.

As far as we know, all these suggestions have not yet been investigated or tried in an adult CHD population. We advocate for more attention for this increasing population and thereby increasing problem. Employers and medical officers or company doctors will increasingly face employees with CHD. Celermayer et al. described a study in 1993 in which employers were questioned on their experiences with CHD. They noticed that in small companies, most decisions about employees with medical problems were made directly by the employers. More important, employers were negative about job prospects for employees with more severe or only partly corrected CHD. To our knowledge, it is unknown how employers nowadays view CHD patients. However, it is impossible to educate all employers. Therefore, education on improved outcome and prognosis of adults with CHD should be widespread among company doctors.

Finally, although beyond the scope of this thesis, the total costs of CHD will go far beyond health-care utilization alone if the loss of work force, caused by the increased unemployment in adult with CHD, is continuing and insufficiently recognized.
REFERENCES


