Cardiol Young 2005; 15: 391–395

ISSN 1047-9511

Original Article

Employment and advice regarding careers for adults with congenital heart disease

David S. Crossland,[1] Susan P. Jackson,[1] Rosalind Lyall,[1] John Burn,[2] John J. O'Sullivan[1]

[1]*Department of Paediatric Cardiology, Freeman Hospital;* [2]*Department of Clinical Genetics, International Centre for Life, Newcastle upon Tyne, United Kingdom*

Abstract *Aims:* To compare the rates of employment, and advice offered concerning careers, in adults with congenital heart disease and controls. To assess the impact of the severity of the congenital cardiac malformation on the chances for employment. *Methods:* We solicited responses from 299 adults with congenitally malformed hearts, asking them to answer questions from a questionnaire posed at interview by a trained nurse. The adults were asked to give an identical questionnaire to a friend to act as a control. We received responses from 177 of the controls. *Results:* The responses showed that 51 of 156 (33 per cent) adults with congenital heart disease were unemployed, and 37 of 151 (25 per cent) had been unemployed for more than a year. This is significantly more than 25 of 156 (16 per cent) matched controls unemployed, and 5 of 151 (3 per cent) controls unemployed for more than a year. Almost one-fifth of the adults with congenital heart disease (19 per cent) had received advice regarding their career which they found helpful, which is significantly fewer than the 31 per cent of controls. More (42 per cent) had been given advice against certain occupations than controls (11 per cent). Receiving career advice was significantly associated with employment in the population with congenitally malformed hearts, with almost three-quarters (73 per cent) of those given advice being employed compared to 46 per cent of those not given advice. This pattern was not seen in controls. The severity of the congenital cardiac malformation did not significantly affect the rates of unemployment. *Conclusions:* Whatever the severity of their disease, adults with congenitally malformed hearts are more likely to be unemployed than matched controls. They are less likely to receive useful advice regarding potential careers, and find the advice given less helpful, than controls, although receiving suitable advice is associated with being employed in the population with congenital cardiac disease.

Keywords: Congenitally malformed hearts; grown-up congenital heart disease; unemployment

THE MAJORITY OF CHILDREN WITH CONGENITALLY malformed hearts now survive into adulthood.[1,2] There has, therefore, been increased interest in the psychosocial functioning, state of health, and quality of life of adults with congenital heart disease.[3–9] The ability to obtain employment is an important factor, influencing both quality of life and social inclusion.

Studies of subgroups of patients with congenital heart disease suggest a similar, or even higher, rate of employment than the general population of their country.[3,8,10] A Finnish study, comparing all those with congenital heart disease with randomly selected controls, found similar rates of employment between the two groups.[11] There are conflicting results as to the influence of the severity of the congenital cardiac malformation on the outcomes of employment, although there is suggestion that there is reduced job participation in those with more complex disease.[3,12,13]

Our study compares the rates of employment of adults being followed up with congenital heart disease with controls, and looks at the impact of the severity of their lesion on the outcome of employment. We also explored the advice regarding careers given to the adults with congenitally malformed hearts.

Correspondence to: David S. Crossland, Department of Paediatric Cardiology, Freeman Hospital, Newcastle upon Tyne NE7 7DN, United Kingdom. Tel: +44 191 213 7146; Fax: +44 191 223 1314; E-mail: davidxland@hotmail.com

Accepted for publication 26 January 2005

Methods

Patients under follow-up with congenitally malformed hearts were selected at random from the departmental database, and sent a letter asking them if they would like to participate in the study. Patients were also recruited at our clinic for adults with congenital heart disease. In total, we approached 320 patients over the age of 16 years with congenital heart disease, and 299 of these agreed to participate. We excluded any patients known to have difficulties with learning. The structured questionnaire was posed in person by one of our research nurses, either in a quiet room in the outpatient department or in the home of the patient. Questions were asked under the categories of education, advice regarding potential careers, employment, and exercise ability. The majority of the questions required a "yes" or "no" answer to be ticked, such as "are you currently employed" and "have you ever been out of work for more than a year"? Questions that required more elaboration were left open. For example, the question "have you ever been advised against a specific occupation?", which required the answer of "yes" or "no", was followed by "who gave you this advice?" Including an explanation of the study, and the time required to obtain consent, the questionnaire took between 20 and 30 minutes to complete.

The patients with congenitally malformed hearts were divided into categories of mild, significant, and complex lesions, attempting to take into account the original diagnosis, previous surgery, and the need for future intervention. Mild disease was classified as those needing observation only, with no prospect of intervention in the foreseeable future. This included lesions such as mild aortic or mitral regurgitation, mild pulmonary stenosis, and those with atrial or ventricular septal defects subsequent to repair. Significant lesions included those in whom a correction has been attempted, but who are likely to require further intervention, or to have further medical problems related to their congenital cardiac malformation. Such patients included those with tetralogy of Fallot, those who had undergone a Ross procedure, those with repaired coarctation, and those with repaired atrioventricular septal defect with common valvar orifice. Complex lesions are those where corrective surgery could not be carried out, including those with the Eisenmenger reaction, those whose pulmonary circulations were shunt-dependant, those with Fontan circulations, and those with the morphologically right ventricle supplying the systemic circulation.

To obtain a control group of similar social background and expectations to the population of patients with congenitally malformed hearts, each adult with congenital heart disease was asked to give an identical questionnaire to a friend without congenital heart disease. Of the prospective controls, 177 posted back their questionnaire. The questionnaire included detailed questions about physical activity so that we were able to calculate accurately the standing within the classification of the New York Heart Association in both groups.

We used McNemar's test to compare matched pairs, and the Chi-squared test to compare proportions. If patients, or either of a matched pair, had not answered a question, they were excluded from the denominator for that question.

Ethics approval was obtained from the Joint Ethics Committee of Newcastle and North Tyneside Health Authority, University of Newcastle upon Tyne, and the University of Northumbria and Newcastle.

Results: all patients with congenital heart disease

Of the 299 patients, 137 were male (46 per cent), and the mean age was 30.9 years, with a range of 17–83 years. Within our categories, 109 (36 per cent) were deemed to have mild disease, 144 (48 per cent) significant disease, and 46 (16 per cent) severe disease. Excluding students, those on maternity leave, and those who had retired, 90 of 273 (33 per cent) were not working at the time of the questionnaire. Of this 273, 66 had been out of work at some point for more than a year. This high rate of unemployment is seen across the spectrum of congenital cardiac malformations, with no significant difference in rates of unemployment according to the severity of the different lesions (Table 1). The rate of employment of men and women with congenitally malformed hearts was the same, with 44 of 132 (33 per cent) men, and 46 of 141 (33 per cent) women, not working at the time of the questionnaire. Women were more likely to be working part time than men, with 31 of 95 (33 per cent) women compared to 7 of 60 (12 per cent) men employed part time, p equal to 0.0011. At the time of the study, the regional rate of unemployment was 8.3 per cent, and the national rate was 6.1 per cent.[14]

Table 1. Unemployment rates at different severities of congenital heart disease.

Severity of congenital heart disease	Unemployed	Out of work for more than 1 year
Mild	32/98 (32%)	19/98 (19%)
Significant	40/133 (30%)	33/133 (25%)
Complex	18/42 (43%)	14/42 (33%)
Chi-squared p	0.57	0.39

Results: comparison of matched pairs

Of those with congenitally malformed hearts whose control replied, 81 of 177 (46 per cent) were male, compared to 69 of 177 (39 per cent) controls. The mean age for the matched group with congenital heart disease was 32.4 years, with a range of 17–83 years, and the mean age of their matched controls was 32.9, with a range of 17–81 years.

Employment

Students, those on maternity leave, and those who had retired, were again excluded from the analysis regarding employment. When compared to matched controls, adults with congenital heart disease were significantly more likely to be out of work, and also more likely to have been out of work for greater than 1 year (Table 2). Although more of those with congenitally malformed hearts had limitations in their capacity to exercise, the difference in rates of employment remains significant when only cases and controls with the same tolerance as judged using the categorisation of the New York Heart Association are compared (Table 2). Among those who were employed, there was no significant difference in the proportions of each group working part time, with 17 of 87 (21 per cent) of those with congenitally malformed hearts working part time, compared to 26 of 87 (30 per cent) controls, p equal to 0.14.

Table 2. Unemployment rates in adults with congenital heart disease and their controls. "Exercise tolerance matched" includes only those matched pairs in which both the patient and control are in the same category within the system devised by the New York Heart Association.

	Unemployed	Unemployed for more than 1 year
All pairs		
Congenital heart disease	51/156 (33%)	37/151 (25%)
Controls	25/156 (16%)	5/151 (3%)
McNemar p	<0.0001	<0.0001
Exercise tolerance matched		
Congenital heart disease	21/96 (22%)	14/96 (15%)
Controls	8/96 (8%)	4/96 (4%)
McNemar p	0.0059	0.0162

Career advice and education

The mean age at leaving school was 16.4 years both for the adults with congenital heart disease and their controls, with a mean difference in age at leaving school between matched pairs of less than 0.1 years. Advice regarding potential careers, whether or not the advice was thought to be helpful, and participation in educational and vocational courses are shown in Table 3. Despite being less likely to consider the advice offered helpful, receipt of advice in the overall group of those with congenitally malformed hearts was associated with being employed, with 157 of 216 (73 per cent) patients who had received advice being employed, compared to 25 of 54 (46 per cent) who had not received advice being employed, p equal to 0.0002. This pattern was not significant in controls, with 122 of 146 (84 per cent) controls who had received advice being employed, compared to 17 of 21 (81 per cent) controls not receiving advice being employed, p equal to 0.09. A greater proportion of the patients with congenitally malformed hearts had been given advice against a specific career. A cardiologist had given this advice in 43 per cent of cases, a career advisor in 24 per cent, and a general practitioner in 11 per cent.

Discussion

To the best of our knowledge, ours is the first large study using matched controls to examine the state of employment of adults with congenital heart disease. We have shown that such adults are significantly more likely to be unemployed than their matched controls. It is of great concern that over a quarter of our patients has been out of work for more than 1 year. It is worth noting that both our patients and their matched controls have high rates of unemployment compared to the regional rate of unemployment. This is due in part to different definitions of unemployment, but it clearly illustrates the importance of having a properly controlled population. The reasons

Table 3. Participation in further courses, and advice given concerning potential careers, in the patients with congenital heart disease and their matched controls. Numbers are patients answering "yes" to each question.

	Congenital heart disease	Controls	McNemar p
Have you done an educational/vocational course?	145/174 (83%)	123/174 (71%)	0.004
Have you ever received career advice?	143/176 (81%)	151/176 (86%)	0.256
Did you find the career advice helpful?	26/134 (19%)	41/134 (31%)	0.037
Have you ever been advised against a specific job?	71/169 (42%)	19/169 (11%)	<0.0001

for this difference in unemployment are not straightforward, particularly given that unemployment was high across the whole spectrum of congenital cardiac malformations, and when taking into account limitations of exercise. The age at leaving school was comparable between the two groups, although we did not ask about academic achievement. Interestingly, more of the patients with congenitally malformed hearts proceeded to further education or a course, suggesting that motivation is not a problem. Given the ultimate outcome for employment, a likely explanation for this is that going onto further training is a reflection of difficulty in finding work, and that the courses were undertaken in an effort to gain employment. In the United States of America, it has been shown that patients with aortic stenosis, pulmonary stenosis, or a ventricular septal defect achieve a higher educational level than do the general population, and subsequently go on to have comparable rates of employment.[10]

The situation with advice regarding potential careers is unsatisfactory. As expected, similar numbers received advice, though fewer of those with congenital heart disease had found the advice helpful. Many were given advice against specific jobs, often by medical staff not trained to balance this with directed positive advice, or by career advisors who are not trained to give advice based on the medical condition of the patient. We have shown that, despite not finding the advice helpful, receiving advice was associated with being employed in those with congenitally malformed hearts. This may not be as a direct result of the advice given, as those who seek advice may also be more likely to obtain employment.

There are many possible ways of trying to improve the states of employment of adults with congenitally malformed hearts. The most obvious is helpful, positive, yet medically appropriate advice concerning potential careers. Guidelines based on each diagnosis, to which the advisor can refer, are far from perfect. This is particularly true for those lesions considered to be significant, as there is a wide variation in haemodynamic outcome within each headline diagnosis.[15–17] Individualised discussion with the patient, involving both the cardiologist and the career advisor, preferably at a single clinic, seems more appropriate. An adolescent or transition clinic, as advised by the intercollegiate working party of the United Kingdom on adolescent health, would seem an ideal forum for such discussion.[18] We found disappointing rates of employment for all severities of disease, and did not see the worse rates of employment in those deemed to have more complex disease, as had been noted by others.[12,13] All our patients were chosen from a group of patients with congenitally malformed hearts under follow up, not a cohort followed from birth, and it may be that the label of follow-up is a disadvantage as far as application for employment is concerned. Educating potential employers about individual patients, seeking to prevent overestimation of the problems associated with mild disease, and underestimation of the difficulties created by complex disease, could help individual applications. Patients themselves may not be able to give a sufficiently accurate summary of their underlying condition, putting off potential employers even if the disease is mild.[19]

Limitations of our study

We have shown a strong correlation between having a congenitally malformed heart and being out of work. Further investigation is required to ascertain the exact reasons for this. We did not ask whether our patients who were unemployed were actively looking for work, whether any had taken early retirement, or how many of the patients, be they employed or unemployed, had told prospective employers about their congenital malformations. Although we assume that the congenital malformation was itself a significant factor in any difficulty in finding work, there may have been other comorbidities influencing the outcome with regard to employment. Further studies are needed to look into whether patients who are working are in the employment of their choice, and whether they experience difficulties with progression of their careers, or other aspects of work such a pension because of their congenitally malformed heart.

The population of adults with congenital heart disease is increasing. The majority does not require intervention, and is surgically cured or corrected. Irrespective of the severity of their disease, this population has a high rate of unemployment, potentially affecting social inclusion and the quality of their life. As a group, they find the advice given about potential careers to be unhelpful. We consider that it is important to discuss and address these issues at clinics designed specifically for adolescents and adults with congenital heart disease so as to make our patients aware of the potential difficulties, and help them find ways to overcome them.

Acknowledgements

Our study was supported by a grant from the Ann Coleman research fund. We thank Mark Pearce for his statistical advice and calculations.

References

1. Wren C, O'Sullivan JJ. Survival with congenital heart disease and need for follow up in adult life. Heart 2001; 85: 438–443.
2. Celermajer DS, Deanfield JE. Adults with congenital heart disease. BMJ 1991; 303: 1413–1414.

3. van Rijen EH, Utens EM, Roos-Hesselink JW, et al. Psychosocial functioning of the adult with congenital heart disease: a 20–33 years follow-up. Eur Heart J 2003; 24: 673–683.
4. Lane DA, Lip GYH, Millane TA. Quality of life in adults with congenital heart disease. Heart 2002; 88: 71–75.
5. Kamphuis M, Ottenkamp J, Vliegen HW, et al. Health related quality of life and health status in adult survivors with previously operated complex congenital heart disease. Heart 2002; 87: 356–362.
6. Saliba Z, Butera G, Bonnet D, et al. Quality of life and perceived health status in surviving adults with univentricular heart. Heart 2001; 86: 69–73.
7. Van Doorn C, Yates R, Tunstill A, Elliot M. Quality of life in children following mitral valve replacement. Heart 2000; 84: 643–647.
8. Ternestedt BM, Wall K, Oddsson H, Riesenfeld T, Groth I, Schollin J. Quality of life 20 and 30 years after surgery in patients operated on for tetralogy of fallot and for atrial septal defect. Pediatr Cardiol 2001; 22: 128–132.
9. Popelova J, Slavik Z, Skovranek J. Are cyanosed adults with congenital cardiac malformations depressed? Cardiol Young 2001; 11: 379–384.
10. Gersony WM, Hayes CJ, Driscoll DJ, et al. Second natural history study of congenital heart defects: quality of life in patients with aortic stenosis, pulmonary stenosis, or ventricular septal defect. Circulation 1993; 87 (1 Suppl): 52–65.
11. Kokkonen J, Paavilainen T. Social adaptation of young adults with congenital heart disease. Int J Cardiol 1992; 36: 23–29.
12. Celermajer DS, Deanfield JE. Employment and insurance for young adults with congenital heart disease. Br Heart J 1993; 69: 539–543.
13. Kamphuis M, Vogels T, Ottenkamp J, Van der Wall EE, Verloove-Vanhorick SP, Vliegen HW. Employment in adults with congenital heart disease. Arch Pediatr Adolesc Med 2002; 156: 1143–1148.
14. Office for National Statistics. Region in figures, North East. Winter 2000, No. 2. Table 4.1.
15. Celermajer DS, Greaves K. Survivors of coarctation repair: fixed but not cured. Heart 2002; 88: 113–114.
16. van Doorn C. The unnatural history of tetralogy of Fallot: surgical repair is not as definitive as previously thought. Heart 2002; 88: 447–448.
17. Deanfield J, Thaulow E, Warnes C, et al. Management of grown up congenital heart disease. Eur Heart J 2003; 24: 1035–1084.
18. Bridging the Gaps: Health Care for Adolescents. The intercollegiate working party on adolescent health. Royal College of Paediatrics and Child Health, June 2003.
19. Kantoch MJ, Collins-Nakai RL, Medwid S, Ungstad E, Taylor DA. Adult patients' knowledge about their congenital heart disease. Can J Cardiol 1997; 13: 641–645.