Cardiol Young 2007; 17: 64–71

ISSN 1047-9511
doi: 10.1017/S1047951106001016

Original Article

Outcome after surgical repair of congenital cardiac malformations at school age

Rachel E.A. van der Rijken,[1] Ben A.M. Maassen,[1] Twiggy L.M. Walk,[2] Otto Daniëls,[2] Gerdine M. Hulstijn-Dirkmaat[1]

[1]*Department of Medical Psychology,* [2]*Children's Heart Centre, Radboud University Nijmegen Medical Centre, Nijmegen, The Netherlands*

Abstract *Objectives:* To explore the long-term physical, educational, behavioural, and emotional outcome of patients undergoing surgical correction of congenital cardiac disease at school age, and to investigate the relation, if any, between the outcome and comorbidity, age and sex, and level of complexity of the cardiac surgery. *Methods:* Information was obtained concerning 101 patients who underwent open-heart surgery for correction of congenital cardiac malformations between 1992 and 2000 whilst aged from 6 to 16 years. The patients, and their parents, completed the questionnaire "Outcome of congenital heart disease and surgery", the RAND 36-Item Health Survey, and the Child Behaviour Checklist/Youth Self-Report/Young Adult Self-Report. *Results:* Of the patients, 26% had comorbidity. Of those without comorbidity, 39% had frequent physical complaints, and 28% experienced limitations due to the cardiac disease. Nevertheless, the patients reported a good subjective state of health, and did not report any behavioural or emotional problems. Patients did show academic difficulties. They had received special education more frequently than their healthy peers, and many had needed to repeat a grade, or had received remedial teaching. Consequently, the educational level of patients was lower than that of their healthy peers. Patients with comorbidity, female patients, and patients who underwent complex surgery, seemed to be most at risk for physical, behavioural, and emotional problems. *Conclusion:* It is necessary to distinguish between physical state and its appraisal, and clinicians should be aware of this. Further research is needed to find out the cause and nature of the academic difficulties. Groups of patients at risk should be followed closely to enable early interventions.

Keywords: Long term outcomes; education; emotional state; behavioural problems

Advances in diagnosis and treatment for congenital disease have resulted in improved outcomes in children born with cardiac malformations. Although the majority of these children seem to be functioning at a normal level,[1] as a group they are at risk of developing educational, behavioural, and emotional problems.[2–5] Problems mostly appear at school age, when children have to meet specific cognitive and social demands. The cardiac disease, and its treatment, may cause absence from school, thereby putting school performance at risk, and limiting social interactions and contacts with peers. Moreover, the illness may cause the parents to be anxious and overprotective towards the child, which may influence the subsequent development.[6]

In addition to the problems reported at school age, problems have been reported in adulthood. Research, however, has yielded contradictory results. Van Rijen et al.[7] reported lower levels of education in patients compared with a healthy population, whereas Nieminen et al.[8] found similar educational levels. Ternestedt et al.[9] showed even higher educational levels in adults with a congenital cardiac malformation compared with healthy controls. With regard to quality of life, Kamphuis et al.[10] found that patients with a complex malformation functioned less well compared with the general population, whereas

Correspondence to: Rachel E.A. van der Rijken, Radboud University Nijmegen Medical Centre, 840 Medical Psychology, P.O. Box 9101, 6500 HB Nijmegen, The Netherlands. Tel: +31 24 3613947; Fax: +31 24 3613425; E-mail: R.vanderRijken@cukz.umcn.nl

Accepted for publication 10 March 2006

Fekkes et al.,[11] and Moons et al.,[12] reported the same or even better functioning in patients compared with healthy controls. Regarding aspects of emotional and social functioning in adults, studies have mainly shown favourable outcomes.[7,13,14]

Thus, both favourable and unfavourable outcomes have been shown in patients with congenital cardiac disease. The discrepancies in the outcomes may be related to the heterogeneity of the populations studied, and the diversity of variables in the research design. Our study focuses on the outcome of patients with congenital cardiac malformations undergoing surgical correction during school age. Although nowadays many patients with a congenital cardiac malformation have undergone one or more operations prior to beginning schooling, cardiac surgery at school age may have a considerable impact on development, in part because of the surgical procedure used, but also because of factors such as absence from school, being different from peers, and parental emotional reactions. To examine the impact of cardiac surgery at school age, we designed a study consisting of two parts. The first part is a longitudinal prospective study aimed at neurocognitive functioning in such patients before their surgery, at discharge from hospital, and one year after surgery. The design allows the establishment of causal relations between the specific surgical procedure undergone and the neurocognitive outcome. Moreover, the design controls for medical history, because the patients function as their own controls.

As the neurocognitive effects of open-heart surgery may result in educational problems and behavioural and emotional problems,[3–5] we wanted to add information about the long-term functioning of the patients studied having undergone open-heart surgery at school age. We designed, therefore, the second part of the study, which we present in this manuscript. The objective was to explore the general long-term physical, educational, behavioural, and emotional outcome of congenital cardiac disease treated by cardiac surgery at school age. In addition to exploring the general outcome, we specifically related the outcome to comorbidity, age and sex, and the level of complexity of the cardiac surgery.

Methods

Patients and procedure

All patients who underwent their latest open-heart operation for congenital cardiac disease at our medical centre over the years 1992 up to and including 2000, and who were aged from 6 to 16 at the time of the surgery, were eligible to participate. To have a sample representative of the total population of patients with a congenital cardiac malformation, we included patients with comorbidity. Of the 152 eligible patients, 4 had died, while 7 were excluded because their current address could not be obtained. The remaining 141 patients, and/or their parents, received a letter requesting informed consent accompanied by a set of questionnaires. Table 1 shows the sets of questionnaires used and the responses required. Patients and parents were asked to return the completed questionnaires within four weeks. In addition to the data provided by the patients and the parents, all medical files were searched for age, sex, comorbidity, type of congenital cardiac disease, and type of cardiac surgery. The study was approved by the Institutional Review Board.

To enable comparisons with the associated study, in which causal relations may be established between the outcome and the surgical procedure undergone, we chose to grade the level of complexity of the surgery rather than the lesion. For all patients, the surgical procedure was classified into one of four groups according to the Aristotle™ classification.[15] We preferred the Aristotle approach[15] over the RACHS system[16] because we wanted to indicate the complexity of the surgical procedure and its associated morbidity rather than the risk of death associated with the surgical procedure. In addition, the nomenclature used in our centre corresponds with the nomenclature of the Aristotle approach, which enables reliable translations of surgical procedures into the level of complexity suggested by the Aristotelian approach.

Patient characteristics

Of the 141 sets of questionnaires sent, 101 were returned (71.6%). Information was obtained about 55 male and 46 female patients, the overall group having a mean age of 18.1 years, with a range from 9.3 to 27.4 years. Of the patients, 7 were younger than 11 years, 46 patients were aged 11 to 18 years, and 48 patients were aged 18 years and older. Patients in the group responding did not differ from those not

Table 1. Sets of questionnaires depending on patient's age.

Patient's age	Respondent	Questionnaire
<18 years	Parent	– "Outcome of congenital heart disease and surgery"[17] – Child Behaviour Checklist/6–18[21]
	Patient (≥11 years)	– RAND 36-Item Health Survey[19] – Youth Self-Report[22]
≥18 years	Patient	– "Outcome of congenital heart disease and surgery"[17] – RAND 36-Item Health Survey[19] – Young Adult Self-Report[24]

https://doi.org/10.1017/S1047951106001016 Published online by Cambridge University Press

responding regarding their demographic and medical data (χ^2 tests and t tests, p more than 0.05).

Just over one-quarter (25.7%) of the patients had comorbidity. Of these, 11 patients had chromosomal or syndromal anomalies, 1 patient had haemophilia, 1 patient had hypercholesterolaemia, 4 patients had defects of the central nervous system, 2 patients had respiratory tract disease, 2 patients had scoliosis, 1 patient had undergone nephrectomy, 1 patient had lost a limb, 1 patient had had cancer, and 2 patients had multiple anomalies. In addition, 9 of the patients with comorbidity were cognitively impaired.

Outcome measures

The questionnaire "Outcome of congenital heart disease and surgery"[17] was designed specifically for the purpose of this study. It focused on the current state of the patient, and measured current physical and educational states.

The Dutch version of the RAND 36-Item Health Survey[18,19] measures the subjective health state by means of eight health concepts. The Dutch version of the Child Behaviour Checklist[20,21] contains questions about behavioural and emotional functioning during the preceding six months as reported by the parents. The answers are transformed into eight syndrome scales. The patients themselves completed self-reports comparable to the Child Behaviour Checklist using the Dutch Youth Self-Report[20,22] and the Dutch Young Adult Self-Report.[23,24]

Statistical analyses

To enable clear interpretation of the outcome being the result of only congenital cardiac disease and cardiac surgery at school age, the results of patients without comorbidity were separated from the results of patients with any type of comorbidity. The results of patients without comorbidity were compared with healthy peers, and with the results of patients with comorbidity. In addition, comparisons were made between patients younger than 18 years and patients older than 18 years, male patients and female patients, and the four groups according to the level of complexity of the surgical procedures. Age, sex, comorbidity, and complexity were not confounded.

The questionnaire "Outcome of congenital heart disease and surgery" was analysed in an explorative manner by means of descriptive statistics. The outcome was examined for all patients together, irrespective of respondent. Binomial tests were used to compare the educational data with data from the general Dutch population.[25,26] Chi-square tests, independent t tests, and analyses of variance were used to compare the results of the different groups.

Results on the RAND 36-Item Health Survey, Child Behaviour Checklist, Youth Self-Report, and Young Adult Self-Report were compared with reference data[20,27] by means of binomial tests. Chi-square tests were used to compare the results of the different patient groups.

On the RAND 36-Item Health Survey, the Youth Self-Report, and the Young Adult Self-Report, some of the patients were not able to complete the questionnaires themselves because of cognitive impairment. Self-reports completed by parents instead of patients were excluded, this being required for 4 patients younger than 18 years, and 5 patients older than 18 years.

All statistical analyses were carried out using a two-tailed probability level of 0.05 using the Statistical Package for the Social Sciences version 11.0 for Windows.

Results

Current physical state

General outcome. As shown in Table 2, 43.6% of all patients reported physical complaints. Almost one third experienced limitations due to the cardiac disease, and nearly one quarter experienced limitations due to another disease. Nevertheless, the value of their current health was 7.7 on a scale from zero to ten.

Comorbidity. Even when patients with comorbidity were excluded, 38.7% of the patients reported physical complaints. Of the patients without comorbidity, 28%

Table 2. Current physical state of patients.

		Comorbidity		Sex		Complexity level of surgery			
	All (N = 101)	No (n = 75)	Yes (n = 26)	Male (n = 55)	Female (n = 46)	1 (n = 18)	2 (n = 28)	3 (n = 33)	4 (n = 22)
Frequent physical complaints	43.6	38.7	57.7	**34.5**	**54.3***	38.9	46.4	39.4	50.0
Limitations due to cardiac disease	31.7	28.0	42.3	29.1	34.8	**11.1**	**17.9**	**45.5**	**45.5***
Limitations due to other disease	23.8	**10.7**	**61.5****	27.3	19.6	22.2	21.4	24.2	27.3
Mean value of current health[a]	7.7	**7.9**	**7.0****	7.8	7.6	7.9	7.5	7.8	7.6

All values are given as percentages. Note that the numbers in bold differ significantly between the groups compared: *$p < 0.05$. **$p < 0.01$
[a]Patients and parents valued current health on a scale from zero to ten (ten being the most positive score)

https://doi.org/10.1017/S1047951106001016 Published online by Cambridge University Press

reported limitations due to the cardiac disease (Table 2). As might be expected, a higher proportion of patients with comorbidity experienced limitations due to another disease than patients without comorbidity (χ^2 equal to 27.58; p less than 0.01). In accordance, patients with comorbidity valued their health as poorer than the other patients (t equal to 3.82; p less than 0.01).

Age and sex. Age was not related to physical complaints or limitations (data not shown). Female patients experienced more physical complaints than male patients (χ^2 equal to 4.00; p less than 0.05).

Level of complexity of the cardiac surgery. More patients who had undergone surgical procedures graded at levels 3 and 4 reported limitations due to the cardiac disease than those undergoing procedures graded at levels 1 and 2 (χ^2 equal to 10.81; p less than 0.05).

Subjective health state

General outcome. In contrast to the amount of physical complaints and limitations reported, patients reported better functioning than healthy peers on 5 of the 8 subscales of the RAND 36-Item Health Survey (Table 3). Regarding the other three subscales, specifically "Physical functioning", "Vitality", and "General health perceptions", the patients and their healthy peers were comparable.

Comorbidity. After exclusion of patients with comorbidity, the results practically remained the same as for the total group (Table 3). Patients with comorbidity scored in the clinical range more often than patients without comorbidity on the scales "Role limitations due to physical health problems" (χ^2 equal to 6.15; p less than 0.05) and "Vitality" (χ^2 equal to 10.06; p less than 0.01).

Age and sex. The effects of age and sex were in accordance with the current physical state reported: Age was not related to subjective health state (data not shown). Female patients reported significantly poorer "Physical functioning" than healthy peers (Table 3). In contrast, male patients showed a better subjective health state than healthy peers on 6 of the 8 subscales. Thus, female patients reported more clinically significant problems than male patients on the subscales "Physical functioning" (χ^2 equal to 11.00; p less than 0.01), "Vitality" (χ^2 equal to 6.89; p less than 0.01), "Bodily pain" (χ^2 equal to 8.02; p less than 0.01), and "General health perceptions" (χ^2 equal to 4.16; p less than 0.05).

Level of complexity of the cardiac surgery. The level of complexity of the surgery (data not shown) did not have any effect on the subjective state of health.

Educational state

General outcome. A considerable proportion of the patients showed poor educational attainments (Table 4). Just over one-quarter (26%) had received special primary education, which was almost 8 times the proportion found in the general Dutch population[26] (binomial test 26.3% versus 3.3%; p less than 0.01). In addition, patients had received lower secondary education, including special secondary education, more often than the reference group (binomial test 75.3% versus 62.5%; p less than 0.05). Accordingly, more patients had followed further education at the vocational level than the reference group (binomial test 78.7% versus 55.6%; p less than 0.01). Finally, more patients than healthy peers[25] had repeated a grade in secondary education (binomial test 20.6% versus 5.0%; p less than 0.01).

Comorbidity. When patients with comorbidity were excluded, we still found a high incidence of special education in patients compared with healthy peers (binomial test 13.3% versus 3.3%; p less than

Table 3. Subjective health state: proportions (%) of scores in the clinically significant range.

		Comorbidity		Sex	
RAND scales	All (N = 83)	No (n = 69)	Yes (n = 14)	Male (n = 46)	Female (n = 37)
Physical functioning	25.6	25.0	28.6	11.1*	43.2*
Social functioning	13.3*	13.0*	14.3	6.5**	21.6
Role limitations physical	14.5*	10.1**	35.7	10.9*	18.9
Role limitations emotional	12.0*	8.7**	28.6	6.5**	18.9
Vitality	24.1	17.4	57.1*	13.0	37.8
Bodily pain	10.8**	11.6*	7.1	2.2**	21.6
General mental health	6.0**	4.3**	14.3	4.3**	8.1*
General health perceptions	26.8	23.5	42.9	17.8	37.8

Note that scores below the 25th percentile of the reference group are in the clinical range.[27] Thus, percentages lower than 25 indicate that less patients than references scored in the clinical range

Only significance levels of patients versus references are shown: *$p < 0.05$. **$p < 0.01$. For significant group differences: see text

https://doi.org/10.1017/S1047951106001016 Published online by Cambridge University Press

Table 4. Educational state of patients.

	All		No comorbidity		Comorbidity		
Education	**N**	*%*	**n**	*%*	**n**	*%*	$\chi^2(1)$
Preschool	*101*		*75*		*26*		
Gone to preschool	81	80.2	64	85.3	17	65.4	4.84*
Primary education[a]	*99*		*75*		*24*		
Special primary education	26	**26.3**	10	**13.3**	16	66.7	26.71**
Regular primary education	73	73.7	65	86.7	8	33.3	26.71**
Repeated a grade in RPE[b]	35	47.9	33	50.8	2	25.0	1.90
Received remedial teaching in RPE	17	23.3	16	24.6	1	12.5	.59
Secondary education[c]	*85*		*64*		*21*		
Special education	17	**20.0**	5	**7.8**	12	57.1	24.05**
Lower general education	47	**55.3**	44	**68.8**	3	14.3	18.97**
Higher and pre-university education	21	24.7	15	23.4	6	28.6	.22
Repeated a grade in RSE[d]	14	**20.6**	11	**18.6**	3	33.3	1.03
Received remedial teaching in RSE	11	16.2	8	13.6	3	33.3	2.25
Further education[e]	*47*		*41*		*6*		
Vocational education	37	**78.7**	34	**82.9**	3	50.0	3.39
Higher and university education	10	21.3	7	17.1	3	50.0	3.39

Note the significance level of patients without comorbidity versus patients with comorbidity: *$p < 0.05$. **$p < 0.01$.
The numbers in bold differ significantly between patients and the Dutch population (see text). Patients with comorbidity were not compared with the general Dutch population
[a]Two patients had not received any education; [b]RPE: regular primary education; [c]Fourteen patients were still in primary school; [d]RSE: regular secondary education; [e]Of the 59 patients at this age level, 12 had not received any further education

0.01). Also the level of secondary and further education remained significantly lower than in the reference group (binomial test 76.6% versus 62.5%; p less than 0.05, binomial test 82.9% versus 55.6%; p less than 0.01). In addition, the incidence of repeating a grade in secondary education (18.6%) was still significantly higher than the 5% in the general Dutch population[25] (p less than 0.01).

Age, sex, and level of complexity of the surgery were not associated with educational state.

Behavioural and emotional functioning

General outcome. Table 5 shows behavioural and emotional problems according to the parents of patients younger than 18 years (Child Behaviour Checklist). The patients showed more "Withdrawn Behaviour", more "Somatic Complaints", and more "Attention Problems" than healthy peers. The results of the self-reports (Youth Self-Report, 11–18 years; Young Adult Self-Report, older than 18 years) are not shown, because the patients themselves did not report any significant problems compared with healthy peers.

Comorbidity. After exclusion of the patients with comorbidity, the parents of patients without comorbidity still reported many "Somatic Complaints" and "Attention Problems" on the Child Behaviour Checklist (Table 5). No significant differences were found between patients with comorbidity and patients without comorbidity for the reports of both parents (Child Behaviour Checklist) and those self-reported (Youth Self-Report and Young Adult Self-Report).

Age and sex. According to the parents, female patients in particular showed more clinically significant problems than healthy peers on several scales of the Child Behaviour Checklist (Table 5). The female patients themselves, however, reported no clinically significant problems (Youth Self-Report and Young Adult Self-Report, data not shown). No significant differences were found between male and female patients on the Child Behaviour Checklist, the Youth Self-Report, and the Young Adult Self-Report.

Level of complexity of the cardiac surgery. As shown in Table 5, parents of patients undergoing procedures graded at level 4 reported most problems in the clinical range. Again, the patients themselves reported no clinically significant problems (data not shown).

Discussion

General outcome

As congenital cardiac disease and cardiac surgery may have a considerable impact on the development of afflicted patients, especially at school age, we designed the present study to explore the long-term physical, educational, behavioural, and emotional outcome. Despite reporting physical complaints and limitations due to the cardiac malformation, the patients rated their subjective health state, as measured by the RAND 36-Item Health Survey, as the same or even better than their healthy peers. This result reveals an intriguing issue with respect to appraisal of chronic conditions. Patients seem to acknowledge the presence

https://doi.org/10.1017/S1047951106001016 Published online by Cambridge University Press

Table 5. Behavioural and emotional functioning: proportions (%) of scores in the clinically significant range.

		Comorbidity		Sex		Complexity level of surgery			
CBCL scales	All (N = 51)	No (n = 38)	Yes (n = 13)	Male (n = 25)	Female (n = 26)	1 (n = 13)	2 (n = 11)	3 (n = 14)	4 (n = 13)
Internalizing problems[a]									
Anxious/Depressed	5.9	2.6	15.4	0.0	11.5**	7.7	9.1	0.0	7.7
Withdrawn behaviour	7.8*	5.3	15.4	4.0	11.5**	7.7	0.0	0.0	23.1**
Somatic complaints	11.8**	10.5**	15.4	4.0	19.2**	0.0	18.2*	0.0	30.8**
Social problems	3.9	2.6	7.7	0.0	7.7	0.0	0.0	0.0	15.4
Thought problems	5.9	2.6	15.4	8.0	3.8	7.7	0.0	0.0	15.4
Attention problems	9.8**	7.9*	15.4	4.0	15.4**	7.7	0.0	0.0	30.8**
Externalizing problems									
Rule-breaking behaviour	0.0	0.0	0.0	0.0	0.0	0.0	0.0	0.0	0.0
Aggressive behaviour	2.0	0.0	7.7	4.0	0.0	0.0	9.1	0.0	0.0

Note that the Child Behaviour Checklist (CBCL) was completed by parents of patients younger than 18 years
Only significance levels of patients versus references are shown: *$p < 0.05$. **$p < 0.01$. For significant group differences: see text
[a]Scores above the 98th percentile of the reference group are in the clinical range[20]

of physical complaints and limitations, but do not negatively appraise them to the extent that they consider their subjective state of health to be affected.

In line with the findings on subjective state of health, the patients tended to answer positively when asked about their appraisal of certain behaviours. They did not report any clinically significant problems on the Youth Self-Report and the Young Adult Self-Report. Problems may, nevertheless, have been present, because the parents reported significant problems on their children on several subscales of the Child Behaviour Checklist. Differences between the presence of problems and their appraisal have also been shown in other chronically ill patients. It seems that, in the majority of patients, a process of psychological adjustment occurs, strengthened by having survived serious threats due to the illness. Most patients, therefore, successfully adapt to the illness and may even report a psychological well-being comparable to that of healthy persons.[28–30]

In addition to the physical, behavioural, and emotional outcome, we explored the educational outcome of the patients. Our findings pointed to more academic difficulties for patients compared with their healthy peers. Even of the patients without comorbidity, a relatively high proportion, about one-sixth, had needed special education. Moreover, a considerable number of patients had repeated a grade or had received remedial teaching. As a result, their educational level was lower than that of the Dutch population.

Previous studies on educational levels in patients with surgically corrected congenital cardiac disease have yielded contradictory results.[7–9] This may be due to the fact that school systems differ between countries, which makes studies difficult to compare, and explains why the results of the present study are only in accordance with the Dutch study by Van Rijen et al.[7] Studies converge, however, with respect to the overall relation between neurocognitive impairment after open heart surgery and educational problems.[2,5] The high incidence of "Attention Problems" on the Child Behaviour Checklist observed in our study also subscribes to this.

Effect of comorbidity, age and sex, and level of complexity of the cardiac surgery

In addition to exploring the general outcome of patients undergoing surgical correction of their cardiac malformation at school age, we explored the effect of comorbidity, age and sex, and the level of complexity of the surgical procedures.

Comorbidity. One-quarter of the patients had comorbidity. But even after exclusion of patients with comorbidity, the results remained stable. The general outcome, therefore, was indeed the result of surgical correction of the malformation at school age, and not an effect of comorbidity. Nevertheless, the presence of comorbidity, including cognitive impairment, added to the problems already present: Patients with comorbidity had more limitations due to another disease, valued their current health as worse, reported a poorer subjective health state, and received special education more frequently than patients without comorbidity. No differences were found between patients with and without comorbidity on the Child Behaviour Checklist, Youth Self-Report, and Young Adult Self-Report. These findings are in accordance with research by Fortin et al.,[31] who showed that patients with comorbidity generally report a poorer quality of life related to the physical domains, but not on the social and psychological domains.

https://doi.org/10.1017/S1047951106001016 Published online by Cambridge University Press

Age and sex. None of the outcome variables were associated with age. The results for male and female patients mainly differed with regard to physical functioning. Female patients experienced more physical complaints than male patients. Accordingly, female patients showed a poorer subjective health state on the physical domains than male patients. These findings are in accordance with previous research.[7,14] Although speculations have been made, no clear explanation for this difference has been given.

Level of complexity of the surgical procedures. The type of surgery the patients had undergone was classified into four levels of complexity.[15] The level was not related to the subsequent educational state of the patients. Patients who had undergone surgical procedures graded at levels 3 and 4 reported more limitations due to the cardiac disease, but their subjective health state was unaffected. Parents of patients undergoing a procedure graded at level 4 reported most behavioural and emotional problems. The patients themselves, however, reported no behavioural and emotional problems.

A discrepancy between the reports of patients and their parents was also shown by Krol et al.[32] and Van Rijen.[33] As suggested earlier, a possible explanation of the difference between the presence of problems as reported by the parents, and their appraisal as reported by the patients, may be the adjustment by the patients to their illness. On the other hand, the perceptions of the parents may have influenced the results. Parents of patients who underwent more complex surgery may have been more anxious and protective, thus reporting more behavioural and emotional problems. Other research has shown that maternal perceptions may indeed influence adjustment of the child.[6] Regardless of the possible explanations, those undergoing the most complex procedures merit especial attention regarding physical limitations and behavioural and emotional problems.

Limitations

We acknowledge some limitations of our present study. The small size of our sample, especially when analysing subgroups, prohibited further classification of the patients into more than two age groups, thereby leaving only a split of above and below 18 years. Meaningful effects of age may have been masked. In addition, the overall group was too small and too diverse to investigate the influence of medical history. We are aware that variables like the number of earlier operations, and the age during earlier operations, may have influenced the outcome, and the perceptions of the illness made by the parents. The main purpose of our study, nonetheless, was to explore the general outcome of surgical correction of congenital cardiac disease at school age, regardless of medical history.

Other limitations are the possible confounding of age and respondent on the one hand, and severity of lesion and complexity of surgery on the other hand. Patients were classified according to the level of complexity of their latest surgical procedure. Patients with very complex lesions whose latest surgery was relatively simple, therefore, may have been graded at lower levels than patients with less severe lesions whose latest surgery was more complex. Although such a categorisation may have masked effects of the level of complexity of the surgery, the differences found between the levels of complexity were in accordance with other research.[10]

Conclusions

Our study has demonstrated that, despite physical complaints and limitations, patients with congenital cardiac disease generally report a good subjective state of health and report no behavioural and emotional problems. Clinicians should be aware, however, that even though patients do not explicitly report them, problems may be present. The explicitness of a question may make a difference. Questions about concrete physical complaints, limitations, and behaviours may reveal problems, whereas questions that ask for subjective interpretation may mask problems.

Another relevant finding for clinicians is that patients seem to have more academic difficulties than their healthy controls. As a consequence, patients are at risk for limitations in their academic and vocational careers. Clinicians should inform themselves about the performance of their patient at school to enable early interventions when problems are reported. Assessment of neurocognitive functioning in children with congenital cardiac disease is needed to determine a profile of strengths and difficulties that will enable adequate interventions. Prospective longitudinal studies are recommended to monitor the effects of medical interventions on neurocognitive development.

References

1. Mahle WT. Neurologic and cognitive outcomes in children with congenital heart disease. Curr Opin Pediatr 2001; 13: 482–486.
2. Bellinger DC, Bernstein JH, Kirkwood MW, Rappaport LA, Newburger JW. Visual-spatial skills in children after open-heart surgery. J Dev Behav Pediatr 2003; 24: 169–179.
3. Samango-Sprouse C, Suddaby EC. Developmental concerns in children with congenital heart disease. Curr Opin Cardiol 1997; 12: 91–98.
4. Utens EMWJ, Verhulst FC, Erdman RAM, Meijboom FJ, Duivenvoorden HJ, Hess J. Psychosociaal functioneren van jeugdigen en jong-volwassenen 9 jaar en langer na operatieve behandeling van een aangeboren hartafwijking op de kinderleeftijd

https://doi.org/10.1017/S1047951106001016 Published online by Cambridge University Press

[Psychosocial functioning of children, adolescents, and young adults nine years and longer after surgical correction for congenital heart disease in childhood]. Ned Tijdschr Geneeskd 1996; 140: 1126–1130.
5. Wright M, Nolan T. Impact of cyanotic heart disease on school performance. Arch Dis Child 1994; 71: 64–70.
6. DeMaso DR, Campis LK, Wypij D, Bertram S, Lipshitz M, Freed M. The impact of maternal perceptions and medical severity on the adjustment of children with congenital heart disease. J Pediatr Psychol 1991; 16: 137–149.
7. Van Rijen EHM, Utens EMWJ, 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.
8. Nieminen H, Sairanen H, Tikanoja T, et al. Long-term results of pediatric cardiac surgery in Finland: Education, employment, marital status, and parenthood. Pediatrics 2003; 112: 1345–1350.
9. 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.
10. 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.
11. Fekkes M, Kamphuis RP, Ottenkamp J, et al. Health-related quality of life in young adults with minor congenital heart disease. Psychol Health 2001; 16: 239–250.
12. Moons P, De Bleser L, Budts W, et al. Health status, functional abilities, and quality of life after the Mustard or Senning operation. Ann Thorac Surg 2004; 77: 1359–1365.
13. Cox D, Lewis G, Stuart G, Murphy K. A cross-sectional study of the prevalence of psychopathology in adults with congenital heart disease. J Psychosom Res 2002; 52: 65–68.
14. Utens EM, Versluis-Den Bieman HJ, Verhulst FC, Meijboom FJ, Erdman RA, Hess J. Psychopathology in young adults with congenital heart disease: Follow-up results. Eur Heart J 1998; 19: 647–651.
15. Lacour-Gayet F. Risk stratification theme for congenital heart surgery. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2002; 5: 148–152.
16. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg 2002; 123: 110–118.
17. Van der Rijken R. Vragenlijst "Status van patiënten met een aangeboren hartafwijking na hartchirurgie" [Questionnaire "Outcome of congenital heart disease and surgery"]. Unpublished questionnaire. Department of Medical Psychology, Radboud University Nijmegen Medical Centre, Nijmegen, The Netherlands, 2003.
18. Hays RD, Sherbourne CD, Mazel RM. The RAND 36-Item Health Survey 1.0. Health Econ 1993; 2: 217–227.
19. Van der Zee KI, Sanderman R. Het meten van de algemene gezondheidstoestand met de RAND-36: een handleiding [Measurement of general health status by means of the RAND-36: A manual]. Noordelijk Centrum voor Gezondheidsvraagstukken, Groningen, The Netherlands, 1993.
20. Achenbach TM, Rescorla LA. Manual for the ASEBA School-Age Forms & Profiles. University of Vermont, Research Center for Children, Youth, & Families, Burlington, 2001.
21. Verhulst FC, Van der Ende J. Gedragsvragenlijst voor kinderen van 6–18 jaar [Child Behaviour Checklist for ages 6–18]. Erasmus Medical Centre – Sophia Children's Hospital, Rotterdam, The Netherlands, 2001.
22. Verhulst FC, Van der Ende J. Zelf in te vullen vragenlijst voor 11–18 jarigen [Youth Self-Report for ages 11–18]. Erasmus Medical Centre – Sophia Children's Hospital, Rotterdam, The Netherlands, 2001.
23. Achenbach TM. Manual for the Young Adult Self-Report and Young Adult Behavior Checklist. University of Vermont, Department of Psychiatry, Burlington, 1997.
24. Verhulst FC. Vragenlijst voor jong-volwassenen van 18–30 jaar [Young Adult Self-Report for ages 18–30]. Erasmus Medical Centre, Rotterdam, The Netherlands, 1997.
25. Statistics Netherlands. Zittenblijvers in het algemeen voortgezet onderwijs [Repeating a grade in regular secondary education]. Kwartaalschrift Onderwijsstatistieken 2000; 7: 33.
26. Statistics Netherlands. Regulier onderwijs: instellingen en leerlingen [Regular education: institutions and students]. Statistics Netherlands, Voorburg/Heerlen, 2003.
27. Rose MS, Koshman ML, Spreng S, Sheldon R. Statistical issues encountered in the comparison of health-related quality of life in diseased patients to published general population norms: Problems and solutions. J Clin Epidemiol 1999; 52: 405–412.
28. Gledhill J, Rangel L, Garralda E. Surviving chronic physical illness: Psychosocial outcome in adult life. Arch Dis Child 2000; 83: 104–110.
29. Noll RB, Gartstein MA, Vannatta K, Correll J, Bukowski WM, Davies WH. Social, emotional, and behavioral functioning of children with cancer. Pediatrics 1999; 103: 71–78.
30. Singer MA, Hopman WM, MacKenzie TA. Physical functioning and mental health in patients with chronic medical conditions. Qual Life Res 1999; 8: 687–691.
31. Fortin M, Lapointe L, Hudon C, Vanasse A, Ntetu AL, Maltais D. Multimorbidity and quality of life in primary care: A systematic review. Health Qual Life Outcomes 2004; 2: 51.
32. Krol Y, Grootenhuis MA, Destrée-Vonk A, Lubbers LJ, Koopman HM, Last BF. Health related quality of life in children with congenital heart disease. Psychol Health 2003; 18: 251–260.
33. Van Rijen EHM. Psychosocial aspects of congenital heart disease in adulthood: A longitudinal cohort study of 20–33 years follow-up [dissertation]. Erasmus Medical Centre, Rotterdam, The Netherlands, 2003.