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Cognitive and behavioural status of paediatric patients 1 year after cardiac or cardiopulmonary transplantation

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Abstract. Cardiac and cardiopulmonary transplantation are being increasingly used as methods of treatment for children with end-stage heart or lung disease, but little is documented about their psychological adjustment to such procedures. In all, 45 children who had undergone heart or heart-lung transplantation were studied 1 year after operation and compared with 39 children who had undergone bone marrow transplantation, 49 children who had undergone conventional cardiac surgery and 46 normal healthy children. None of the 3 treatment groups differed significantly with respect to developmental, cognitive or behavioural measures, but there were significant differences between the transplant and normal groups in the areas of developmental and cognitive functioning. The initial diagnosis of the transplant patients was found to be an important factor in the postoperative psychological functioning.

Key words: Behaviour – Cognition – Heart Transplantation – Heart-lung transplantation – Children

With the increasing use of cardiac and cardiopulmonary transplantation for the treatment of children with end-stage heart or lung disease, attention is now being given to the psychological adjustment of children following these procedures. Although it is accepted that children with chronic illness are more likely to experience major psychological and social difficulties than their healthy peers [18], there are still very few published data on the psychological aspects of paediatric heart and heart-lung transplantation. Studies of children 1–5 years after heart transplantation suggested that in most cases the children had returned to age-appropriate activities, including school, and that few were experiencing cardiac-related symptoms [4, 10, 16, 17]. Whilst the side effects of immunosuppressive therapy are still not clear, poor growth has been found in many children taking steroids [24], which may have significant psychological repercussions. The effects of transplantation on the parents have also been assessed, but these studies, together with those on the children themselves, tend to be based on very small sample sizes. The focus has been on heart, rather than heart-lung, recipients in the school-age group, with little attention being given to preschoolers.

In order to address some of these issues with the Harefield patients, a systematic evaluation of paediatric patients and their families is carried out before and at regular intervals after transplantation. This paper reports some of the findings of patients seen 1 year after heart or heart-lung transplantation.

Methods

Patient selection. Between January 1984 and August 1991, 201 heart and heart-lung transplants were performed at the Harefield Hospital on children and adolescents under the age of 17 years. Of the 63 patients eligible for inclusion in the study (criteria for inclusion were that the child and family spoke English, that they were domiciled in the UK or Eire, that the child was under 17 years of age and that they attended the Harefield Hospital for follow-up), 45 patients have been assessed 1 year after transplantation. (The remaining 18 patients were less than 1 year post-transplant).

There were 19 boys and 26 girls, and 37 (82%) came from intact, 2-parent families. The mean age at assessment was 9.6 years (range 1.3–16.0). Of the 45 patients seen, 29 had undergone heart transplantation, of whom 25 had an original diagnosis of cardiomyopathy, 3 had congenital heart disease, and 1 had Kawasaki disease. Sixteen children had had a heart-lung transplant, 8 of whom had pulmonary vascular disease, and 8 had cystic fibrosis. Twelve (27%) of the patients had undergone previous cardiothoracic surgery related to their disease – primarily those with an initial diagnosis of congenital heart disease. One heart-lung recipient had undergone a second transplant for chronic rejection, and a further 2 patients were awaiting retransplantation. At the time of the assessment, 2 children were hospitalised, and a further 6 patients were experiencing problems with repeated infection and/or rejection episodes. Seven of these 8 patients were heart-lung recipients.

Reference group selection. For comparison, 3 reference groups were assessed with the same measures. The first group consisted of 39 children who had undergone bone marrow transplantation (BMT) for a variety of congenital and acquired disorders (mean age at assessment 8.6 years). The second group consisted of 49 patients

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who had undergone conventional cardiac surgery (mean age at assessment 6.8 years) for correction ($n = 46$) or palliation ($n = 3$) of congenital heart disease. The BMT and cardiac surgery groups were seen 1 year after treatment. The third group was a group of 46 normal healthy children (mean age at assessment 8.2 years). The four groups did not differ significantly with respect to sex and social class, but they differed significantly with respect to age – the cardiac surgery group were significantly younger than the transplant group. The reference groups were selected from a prospective study undertaken at the Westminster Children's Hospital between 1984 and 1988. At the time of assessment a similar proportion of children in the transplant, BMT, and cardiac surgery groups were experiencing medical problems.

Measures used

Cognitive development was assessed using either the Ruth Griffiths Mental Development Scales for children of 0–3.5 years [6] or the short form IQ estimate of the British Ability Scales (BAS) for children of 3.6–17.0 years [3]. Two children in the transplant group could not be tested due to neurological damage following transplantation, which was sustained as a result of cardiac arrest prior to establishing cardiopulmonary bypass. These two children were assessed as being in the severely mentally handicapped range. Two children in the cardiac surgery group also could not be tested due to neurological damage following open heart surgery. Although no measure of cognition or behaviour in these neurologically impaired patients is included in the final analysis, they are mentioned so that the groups studied are representative of the status of patients after transplantation or open heart surgery. School attainment tests covering arithmetic (BAS), reading (BAS) and spelling (Schonell graded spelling test) [23] skills were administered to the children aged 5.0–14.4 years. With the exception of the children with impaired neurological status, all of the children in each of the 3 groups were assessed with the appropriate measures of developmental or cognitive ability.

The behaviour of the children was assessed by questionnaires completed by the parents and teachers. Questionnaire assessment of the behaviour of children under 3 years of age was not undertaken because of the lack of suitable measures. The behaviour at home of children of 3.0–5.0 years was assessed with the Richman BCL [20] and for children of 5.0–17.0 years with the Rutter A scales [22]. The behaviour at school was assessed with the Rutter B scales [22], completed by the teachers. The recommended cut-off scores were used in order to identify those children with a significant degree of problem behaviour. Compliance rates for the completion of the questionnaires were 93% for the transplant group, 82% for the BMT group, 96% for the cardiac surgery group and 93% for the normal group.

In addition to assessing the child, the parents were seen for a semistructured interview during which information was collected on demographic, social and medical variables. Specific questions were asked regarding the amount of schooling missed and the child's previous medical history. The cardiologists at the Harefield Hospital and the hospitals at which the cardiac surgery group were patients were also asked to rate the child's medical condition at the time of the assessment.

Statistical analysis. The developmental and cognitive measures were compared using analysis of variance. The Richman and Rutter scores were analysed using the Kruskal-Wallis analysis of variance for nonparametric data.

Results

Development: 0–3.5 years

Ten children in the transplant group, all of whom had an initial diagnosis of cardiomyopathy, were assessed with the Ruth Griffiths Scales. All of the mean subtest scores and overall IQ score were within the normal range. Comparison with the children in the BMT ($n = 8$), cardiac surgery ($n = 14$) and normal ($n = 11$) groups indicated that the transplant group obtained significantly lower scores in all areas of development compared with the normal group, but none of the groups of children who had undergone medical treatment differed from one another (Fig. 1).

Cognitive ability: 3.6–17.0 years

The performance on all of the subtests and the overall IQ of the transplant patients fell within the normal range. The IQ score for the group ($n = 31$) was 99.94 + 16.77 (SD), and the mean scores on the attainments were arithmetic 92.21 + 18.94, reading 94.42 + 18.13 and spelling 84.37 + 18.55. The initial diagnosis of the transplant patients was categorised into cardiomyopathy ($n = 15$), cystic fibrosis ($n = 8$) and congenital heart disease ($n = 8$). In terms of overall IQ and attainment scores with the initial diagnosis as the dependent variable, children with an initial diagnosis of congenital heart disease performed at a lower level on all of the cognitive parameters. The difference reached significance on the overall IQ score, with both the cardiomyopathy and cystic fibrosis groups obtaining a significantly higher score than the congenital heart disease group (Fig. 2).

Comparison with the reference groups indicated that the transplant group performed at a lower level on all parameters, although the differences only reached statistical significance on the spelling test (Fig. 3).

Behaviour at home

Six of the 27 transplant patients (22.2%) on whom Rutter A data were available obtained scores indicative of a significant degree of problem behaviour at home, and the majority of the behaviour problems were of a neurotic nature. There were again marked differences in the prevalence of problem behaviour between the subgroups.

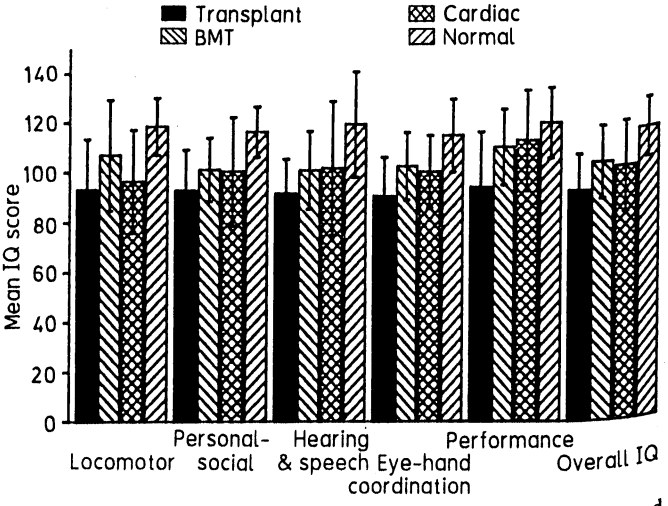

Fig. 1. Developmental ability: comparison of the transplant and reference groups

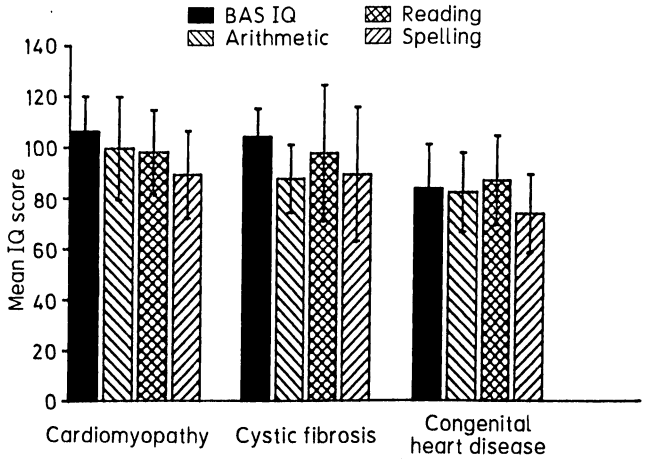

Fig. 2. Cognitive ability of the transplant group according to initial diagnosis

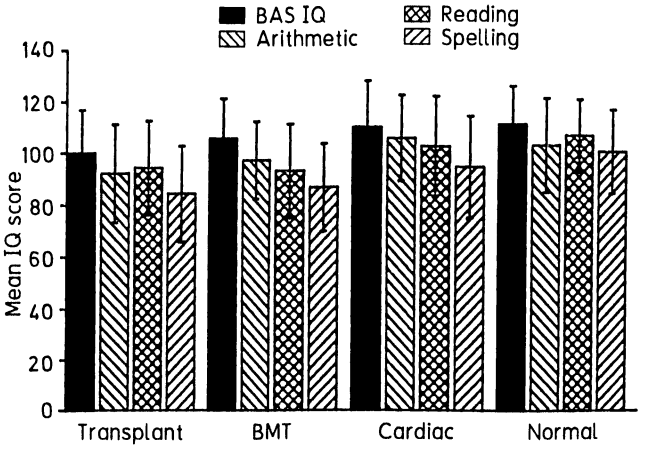

Fig. 3. Comparison of the transplant group with the reference groups: cognitive ability 5–17 years

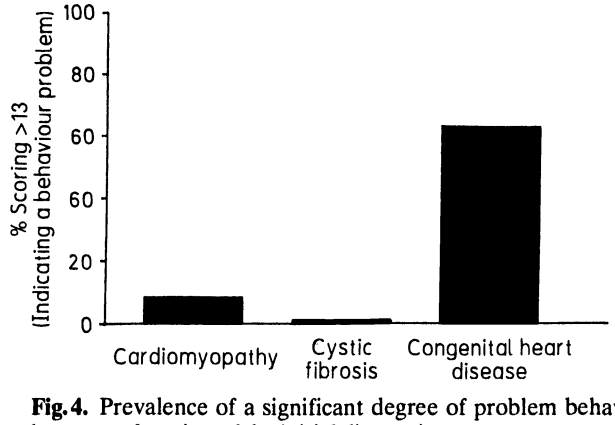

Fig. 4. Prevalence of a significant degree of problem behaviour at home as a function of the initial diagnosis

The group with an initial diagnosis of congenital heart disease obtained a significantly higher score on the Rutter A scale and had a significantly higher proportion of children with scores indicative of problem behaviour than the children originally diagnosed with cardiomyopathy or cystic fibrosis (Fig. 4).

Comparison with the reference groups yielded no statistically significant differences in the prevalence of behaviour problems, although the proportion of children with problem behaviour was higher in the 3 treatment groups than in the normal group (Fig. 5).

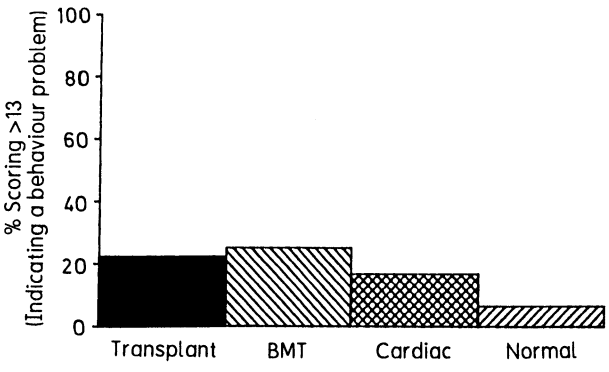

Fig. 5. Comparison of the transplant group with the reference groups: behaviour at home

Behaviour at school

Thirty-two children were eligible for school, 19 of whom were at full-time normal school, 2 were at school part-time, and 3 were at a special school. The reason for non-attendance at school was ongoing medical problems in all 8 cases. One child attending scholl on a part-time basis was able to attend full-time but for psychological reasons was not doing so. This child had had a good medical outcome from the transplantation but had experienced severe adjustment problems posttransplant.

Four of the 18 patients on whom Rutter B data were available were rated as having a significant degree of problem behaviour, of whom 3 children had an original diagnosis of congenital heart disease. Comparison with the reference groups indicated that the prevalence of problem behaviour was similar in all of the groups of children who had undergone medical treatment but was higher than that seen in the normal group.

Discussion

Heart and heart-lung transplantation are major surgical procedures necessitating intensive medical support and follow-up. The risks involved in the treatment are such that transplantation is only undertaken when there is no alternative form of treatment available. The patients are often chronically ill prior to surgery, with a very poor quality of life and life expectancy. Although there is a dramatic improvement in the clinical condition of most patients after transplantation, regular clinic visits are necessary, and uncertainty about the future remains. Both the patient and family have to adjust to the fact that the child is no longer chronically sick and in many cases is able to lead a completely normal life, sometimes for the first time.

The results of this study indicate that the cognitive and behavioural status of transplant patients 1 year after transplantation is similar to that of other groups of children who have, or have had, a chronic illness. The limita-

tions of the data with respect to sample size are acknowledged, but in the context of the paucity of documented research on the psychological adjustment of paediatric heart and heart-lung transplant patients, these findings do address some of the deficits in the existing literature.

The findings in the younger age group suggest that children undergoing cardiac transplantation before the age of 3 years are developing within the normal range, according to the standardised means for the tests, although their development is significantly behind that of a group of normal healthy children. The lower scores obtained by all 3 groups of children who had undergone medical treatment compared with the normal group correspond with findings in the literature that young children suffering from chronic illness, and particularly cardiac-related problems, are at risk for exhibiting developmental delay [11–13, 21].

In the absence of neurological damage, cognitive development in school-age children who have undergone heart or heart-lung transplantation is within the normal range, although the performance was at a lower level compared with the reference groups, particularly on the short-term memory test. Whilst the heart/heart-lung, BMT and cardiac surgery groups were comparable in terms of the level of medical disability at the time of assessment, the initial diagnosis appeared to be an important factor in the postoperative cognitive status, which would explain the trend for lower scores in the transplant group compared with the reference groups. Whilst it is recognised that congenital heart disease can have a deleterious effect on cognitive function, impairment is more frequently associated with the presence of a cyanotic lesion [1, 9, 14, 15]. The cardiac surgery group predominantly consisted of children with acyanotic lesions, whereas the transplant group consisted of a higher proportion of children with cyanotic heart disease. Cystic fibrosis and cardiomyopathy, together with the conditions for which bone marrow transplantation was the chosen treatment, are not known to have any deleterious effect on brain development, and this is substantiated by the performance of the subgroups of the transplant group. In this paper no preoperative measures to indicate whether the IQ score was lower prior to transplantation are available, but other studies of chronically ill children have found this to be the case [19]. The transplant and BMT patients had also missed more schooling than the children in the cardiac surgery and normal groups, which is likely to have contributed to the lower scores obtained by the transplant and BMT groups on the academic attainments.

In terms of the behaviour at home, the number of children on whom Richman BCL data were available was too small for statistical analysis to be valid, but behaviour problems were reported in the preschool age group following transplantation. Temper tantrums, eating and sleeping difficulties and anxieties were areas of specific concern to the parents of all 3 groups of hospitalised children, and the frequency of reporting of such problems was higher than in the normal group.

For the children of 5 years and older, the Rutter A data indicated that a higher proportion of children had a significant degree of problem behaviour than the expected rate of 10 % for the normal population [22]. The types of problems were predominantly of a neurotic nature and included misery, irritabilty, sleeping and eating difficulties and anxieties. The initial diagnosis was again a significant factor in the prevalence of problem behaviour, with children transplanted for congenital heart disease manifesting the greatest degree of problem behaviour. The majority of children obtaining a score indicative of a significant degree of problem behaviour were well at the time of the assessment but were having difficulties in adapting to the transplant and to the changes in their lifestyle now that they were no longer chronically ill. For 3 of the patients in particular, all of whom originally had congenital heart disease, their problems in behaviour were related to a poor body image and a low self-esteem.

The results of the transplant and reference groups, whilst not demonstrating any significant differences between the groups, are relevant in terms of chronic illness/hospitalisation per se. All 3 groups of patients who had undergone medical treatment had a higher prevalence of problem behaviour than their healthy peers, and this corresponds to the level of emotional and behavioural disorder found in other studies of chronically ill children [5, 7]. Increased levels of depression, anxiety and aggression have also been reported in paediatric kidney and bone marrow transplant recipients [2, 8, 19].

The pattern of behaviour at school was very similar to that reported at home, with the children initially diagnosed with congenital heart disease manifesting the highest proportion of problem behaviour. Comparison with the reference groups indicated similar patterns of problem behaviour in the 3 groups of children who had undergone medical treatment, and a higher proportion of children in these 3 groups manifested a significant degree of problem behaviour at school compared with the normal group. The types of problems included poor concentration, lack of motivation and overly anxious behaviour.

This paper reports a single, retrospective assessment of 45 patients 1 year after heart or heart-lung transplantation and clearly indicates that there are important psychological effects associated with such procedures in children. A similar conclusion has also been reached by Uzark and Crowley [25]. The findings indicate the importance of conducting a controlled, prospective study involving all family members, thereby enabling changes over time in psychosocial functioning to be monitored and comparisons to be made with other groups of chronically ill children and also with normal healthy children. Ultimately, therapeutic interventions will be planned to reduce psychiatric morbidity in this group of paediatric patients and families.

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