



## STUDENT PAPER

## Is cardiac surgery warranted in children with Down syndrome?

### A case-controlled review

M A Roussot, J B Lawrenson, J Hewitson, R Smart, H P de Decker

**Objectives.** To compare children with Down syndrome and children without Down syndrome and investigate whether there is a significant difference in the burden that is placed on the health care system between these two groups only in respect of the repair of congenital heart disease at Red Cross War Memorial Children's Hospital, Cape Town, South Africa.

**Design.** This study is a retrospective case control review.

**Setting.** Red Cross War Memorial Children's Hospital, Cape Town, South Africa.

**Subjects.** The sample group of 50 Down syndrome children who had received cardiac surgery between January 1998 and June 2003 was compared with a control group of 50 non-syndromic children who had received cardiac surgery during the same period.

**Outcome measures.** Sex and diagnoses (cardiac and non-cardiac), number of days spent in hospital and in ICU,

complication rates, re-operation rates, early mortality rates, planned further cardiac surgery. Costs of these outcomes were not quantified in exact monetary terms.

**Results.** There was no significant difference between the two groups in terms of the burden that was placed on the health care system. Similar complication rates, re-operation rates and early mortality rates were recorded for both groups. The Down syndrome group appeared to benefit more from cardiac surgery than the non-Down syndrome group.

**Conclusion.** Denying cardiac surgery to children with Down syndrome does not improve the efficiency of resource allocation. It is therefore not reasonable to suggest that the problem of scarce resources can be ameliorated by discriminating against children with Down syndrome.

*S Afr Med J* 2006; 96: 924-930.

Despite an increasing level of skill and technological capacity to provide health care, public health care systems worldwide are being placed under mounting pressure to deliver essential health care with limited resources, South Africa being no exception.<sup>1</sup>

Approximately 40 - 50%<sup>2-4</sup> of children with Down syndrome have congenital heart defects that may be corrected surgically. Down syndrome children are at risk of being discriminated against in the allocation of scarce resources as it may be perceived that they place a greater burden on the public health care system and are less likely to have a favourable outcome to surgical treatment than non-Down syndrome children. A belief exists that the contribution that a Down syndrome

child makes in society, perceived as less significant, does not warrant the child's utilisation of scarce resources. This is best illustrated by the well-publicised case of Baby Ronnie, who was denied surgical repair of tetralogy of Fallot by the Gauteng Department of Health in 2001 on the basis of having Down syndrome.<sup>5</sup> Down syndrome children are relatively easy to identify clinically, and diagnosis can be confirmed by chromosomal analysis, which may predispose these children to being the target of discrimination in the allocation of scarce resources.<sup>6</sup>

The ethical position regarding treatment strategies for Down syndrome children with congenital heart disease (CHD) is debated internationally,<sup>7</sup> and there is evidence that more institutions worldwide are offering the same treatment methods for CHD to children with and without Down syndrome.<sup>8</sup> For details of the dialectic about whether or not to offer surgical correction of CHD to children with Down syndrome in South Africa please refer to Lawrenson *et al.*<sup>6</sup>

The lack of a national policy in South Africa to govern the treatment of Down syndrome children who have CHD has led to diverse treatment strategies in different provinces. As yet, there appears to be insufficient evidence relevant to the South African context on which to base policy decisions.

*Division of Critical Care and Children's Heart Diseases, School of Child and Adolescent Health, University of Cape Town and Red Cross War Memorial Children's Hospital, Rondebosch, Cape Town*

M A Roussot, 5th-year medical student  
J B Lawrenson, FC Paed (SA), MMed  
J Hewitson, MB ChB, FCS (SA) (Cardiothoracics)  
R Smart, BSc (Med) Hons, Dip Med Tech

*Division of Human Genetics, Department of Clinical Laboratory Sciences, Faculty of Health Sciences, University of Cape Town.*

H P de Decker, MSc, FCPaed (SA), Cert Med Genetics (SA)

*Corresponding author: H P De Decker (rd@cormack.uct.ac.za)*



Currently, in South Africa, Red Cross War Memorial Children's Hospital (RXH) in Cape Town offers surgical treatment to Down syndrome children with CHD. At this public hospital, it is believed that there is no significant reason to discriminate against these children. However, some specialist public institutions in South Africa that are in a position to provide surgical care for Down syndrome children with CHD do not follow this policy.

This study compares the burden that Down syndrome children place on health resources with that placed by non-Down syndrome children in respect of palliation or correction of CHD at RXH. It is not a health economic analysis and does not attempt to address the overall burden placed on health care resources by children with Down syndrome. This is beyond the scope of our review. To do so would require life-long economic analyses of cohorts of children with Down syndrome who have and *have not* undergone cardiac surgery. At RXH we do not have access to the latter cohort, since all our Down syndrome children are offered surgery and are only excluded on the grounds of cardiological criteria rendering them unsuitable for palliative or corrective surgery. Rather, our review quantifies the burden (in terms of the parameters outlined below) on cohorts of children with and without Down syndrome who have undergone cardiac surgery at RXH over a 5-year period. Inferences will be drawn from this experience.

## Methods

### Selection of subjects

A list of children who had undergone cardiac surgery at RXH from January 1998 to June 2003 was generated and sorted according to the date of surgery. The RXH Intensive Care Unit (ICU) and the University of Cape Town (UCT) Cytogenetics Laboratory databases were used to identify the sample group of 50 Down syndrome children (whose files were available for review). The control group of 50 children was created from the same cardiac surgery list by selecting the non-Down syndrome child on the list following immediately after each Down syndrome child in the sample group. Any child that was clinically identifiable as having a syndromic disease (e.g. Williams syndrome, 22q deletion syndrome) was excluded from the study. This exclusion was necessary in order to draw a comparison between Down syndrome children and children that were not clinically syndromic.

This method of selection was chosen to minimise the impact that variable medical staff and facilities over the 5-year period would have had on the management of the patients. There was no intentional matching of Down syndrome children with non-Down syndrome children according to the type of cardiac defect, as an impractically larger number of subjects would have had to be reviewed in order to make such a comparison owing to the variance of cardiac lesions between

the two groups. Attempts to extend the study to include children operated on in 1996 and 1997 failed owing to the poor availability of patient information. There was also no intentional matching of samples according to the type of cardiac surgical procedure for the same reason. However, it is important to note that at other institutions Down syndrome children are denied consideration for cardiac surgery irrespective of the type of defect present or procedure required.

Patients' medical files were retrospectively reviewed. Case notes were used to determine patient details such as age, sex, diagnoses (cardiac and non-cardiac), dates of admission and discharge (to and from hospital and ICU), surgical details and mortality. Late mortality rates were not determined because of resource constraints. Data regarding admissions to other hospitals were not included in this study. No other state hospital in the Western or Eastern Cape offers a comprehensive cardiology and cardiac surgery service to children.

### Measurements

In order to evaluate the burden to the public health care system, the groups were compared in terms of the number of days spent in a general hospital ward and in the ICU.

Benefit derived from cardiac surgery was evaluated by determining the number of days spent in RXH (only) before the first cardiac surgical procedure, between procedures (for children who received more than one cardiac surgical procedure) and after final palliation. Since some children did not undergo their final palliation during the study period, any children requiring anticipated further cardiac surgery were enumerated. Children who had been recorded as deceased were excluded from this enumeration.

The postoperative course was evaluated by determining the frequency of re-operations, complications of surgery and rate of early mortality for both groups. Early mortality is defined as death after cardiac surgery prior to discharge. Resource constraints prevented the determination of late mortality rates and prolonged follow-up at hospitals other than RXH.

Admissions for investigative purposes only (e.g. cardiac catheterisation) were included in the number of admissions before and after surgery. This allowed a comparison of hospital resources utilised by both groups even though these admissions represent the health professional's request for the patient's admission and not necessarily the patient's need for admission as a result of illness.

### Data management and analysis

Data were recorded in an MS Access database designed specifically for this study. MS Excel and Stata v.8 were used to process, summarise and present data. The chi-squared test was used to test for statistical associations between categorical variables; however, the Fisher's exact test was used instead



of the chi-squared test in the event of a cell size less than 5. Numerical variables were compared using the two-sample Wilcoxon rank-sum test and the Wilcoxon signed-rank test where appropriate and are given as mean, median, standard deviation (SD) and range.

Results

Sex

The Down syndrome group consisted of 31 females (62%) and 19 males (38%). The non-Down syndrome group consisted of 28 females (56%) and 22 males (44%), p = 0.542.

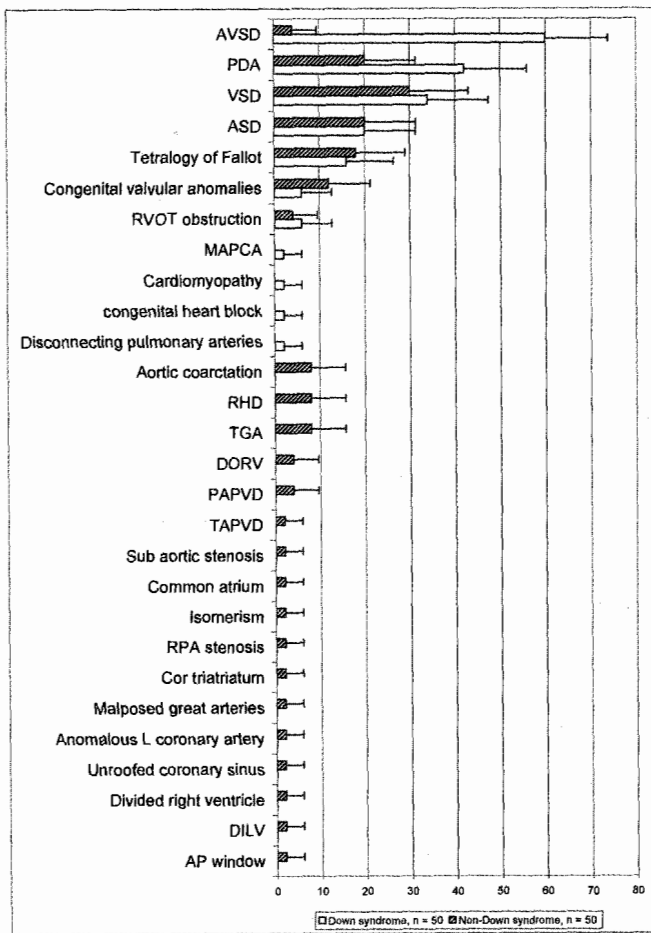


Fig. 1. Distribution of cardiac diagnoses in cohorts of Down syndrome and non-Down syndrome children who have undergone cardiac surgery at Red Cross War Memorial Children's Hospital between January 1998 and June 2003. AVSD - atrioventricular septal defect; ASD - atrial septal defect; VSD - ventricular septal defect; PDA - patent ductus arteriosus; RVOT obstruction - right ventricular outflow tract obstruction; MAPCA - multiple aorto-pulmonary connecting arteries; RHD - rheumatic heart disease; TGA - transposition of the great arteries; DORV - double outlet right ventricle; PAPVD - partial anomalous pulmonary venous drainage; TAPVD - total anomalous pulmonary venous drainage; RPA - right pulmonary artery; DILV - double inlet left ventricle. Error bars represent 2 standard deviations.

Cardiac diagnoses

The distribution of cardiac defects in the Down syndrome group is markedly different to that in the non-Down syndrome group. Fig. 1 illustrates the observed frequency of all cardiac diagnoses, which are not necessarily isolated diagnoses. The Down syndrome group showed less variance than the non-Down syndrome group in terms of the type of cardiac defects present with 11 and 24 different cardiac diagnoses recorded respectively. Atrioventricular septal defects (AVSDs, p < 0.0005) and patent ductus arteriosus (PDA, p = 0.017) were observed to be significantly more common in the Down syndrome group than in the non-Down syndrome group. Ventricular septal defects (VSDs), atrial septal defects (ASDs) and tetralogy of Fallot were common and were similarly distributed in the two groups. Other notable differences in the diagnoses shown in Fig. 1 include transposition of the great arteries (TGA), rheumatic heart disease (RHD) and aortic coarctation, which were each observed with a frequency of 8% in the non-Down syndrome group, but were absent in the Down syndrome group (Fisher's exact test p = 0.117, 1-sided Fisher's exact test p = 0.059).

Non-cardiac diagnoses

A total of 182 non-cardiac diagnoses were recorded for the two groups. The Down syndrome group contributed 105 diagnoses (57.7%) while the non-Down syndrome group contributed 77 diagnoses (42.3%) to this total. The two groups once again showed variance in the distribution of these diagnoses (Fig. 2). Notably, respiratory tract infections (RTIs) were observed in 40 Down syndrome children (80%) and 28 non-Down syndrome children (56%, p = 0.010). Gastroenteritis or diarrhoea was observed in 17 Down syndrome children (34%) and 9 non-Down syndrome children (18%, p = 0.068). Other non-cardiac diagnoses recorded less frequently that are worth highlighting

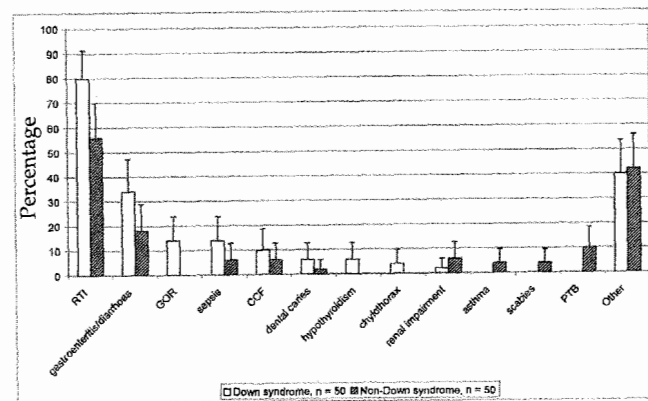


Fig. 2. Non-cardiac diagnoses in cohorts of Down syndrome and non-Down syndrome children who underwent cardiac surgery at Red Cross War Memorial Children's Hospital between January 1998 and June 2003. RTI - respiratory tract infection; GOR - gastro-oesophageal reflux; CCF - congestive cardiac failure; PTB - pulmonary tuberculosis. Error bars represent 2 standard deviations.



include gastro-oesophageal reflux (GOR) recorded in 7 Down syndrome children (14%) but absent in the non-Down syndrome group ( $p = 0.012$ ), and pulmonary tuberculosis (PTB) recorded in 5 non-Down syndrome children (10%) but absent in the Down syndrome group ( $p = 0.056$ ). No significant difference was recorded between the two groups in respect of any of the remaining non-cardiac diagnoses.

### Burden to the health system

The Down syndrome group tended to spend more time in general wards (2 004 days) and in the ICU (437 days) than the non-Down syndrome group (1 420 days and 344 days, respectively). However, there is no significant difference between the two groups in this regard (Table I). The Down syndrome group displayed a greater variance in the time spent in hospital per patient. This is evident in the greater range of days spent in hospital, the general wards and ICU by the Down syndrome group in comparison with the non-Down syndrome group (Table I). The total time spent in hospital for cardiac surgery, the general ward preparation and recovery time as well as the ICU recovery time for the two groups was remarkably similar (Table II).

### Benefit of surgery

The age at first surgery for the Down syndrome group (median: 9.5 months; range: 102.8 months) was significantly less ( $p < 0.05$ ) than the non-Down syndrome group (median: 15.6 months; range 178.3 months). Nevertheless, the Down syndrome group spent significantly more time in hospital than

the non-Down syndrome group *before* surgical intervention. The two groups spent a similar amount of time in the hospital between cardiac surgical procedures, as well as after cardiac surgery (Table III). The Down syndrome group showed a significant reduction in the mean number of days spent in hospital per patient after having had cardiac surgery,  $p = 0.014$  (Fig. 3). In contrast, the non-Down syndrome group did *not* show a significant reduction,  $p = 0.0843$ .

In the Down syndrome group, 7 children (14%) were expected to return for planned further cardiac surgery, while in the non-Down syndrome group, only 4 children (8%,  $p = 0.338$ ) were expected to do so (Fig. 4).

### Postoperative course

The Down syndrome group displayed a lower postoperative complication rate ( $p = 0.205$ ), re-operation rate (Fisher's exact test  $p = 1$ , 1-sided Fisher's exact test  $p = 0.5$ ) and early mortality rate ( $p = 0.538$ ) than the non-Down syndrome children (Fig. 4); however, these differences are not significant. The distribution of postoperative complications was dissimilar for the two groups; the most common postoperative complications in both groups were RTIs, pericardial effusions and sepsis (Fig. 5). The differences in the frequencies of these complications were not significant.

### Discussion

There is substantial evidence to show that the survival profile of Down syndrome children worldwide has been improving remarkably over the past 5 decades,<sup>6-10</sup> although the average

**Table I. Comparison of total days spent in hospital and ICU for any reason**

	Days in hospital			Days in general ward			Days in ICU			Percentage of days in ICU/ days in hospital		
	DS	Non-DS	<i>p</i>	DS	Non-DS	<i>p</i>	DS	Non-DS	<i>p</i>	DS	Non-DS	<i>p</i>
Mean	48.82	35.28		40.08	28.4		8.74	6.88		21.78	22.1	
Median	36.5	30.5	0.085	31	21.5	0.110	5	4	0.141	15.65	17.29	0.858
SD	39.19	26.48		33.77	23.27		10.75	7.9		19.04	18.34	
Range	165	120		146	110		60	20		82.12	78.04	

DS = Down syndrome; ICU = intensive care unit; SD = standard deviation.

**Table II. Number of days spent in hospital in preparation of and recovery from cardiac surgery**

	Total time in hospital for cardiac surgery (days)			GW preparation for cardiac surgery (days)			ICU recovery from cardiac surgery (days)			GW recovery from cardiac surgery (days)		
	DS	Non-DS	<i>p</i>	DS	Non-DS	<i>p</i>	DS	Non-DS	<i>p</i>	DS	Non-DS	<i>p</i>
Mean	29.72	23.72		16.86	11.22		6.54	6.2		12.86	12.5	
Median	15.5	17.5	0.707	6	5	0.408	4.5	4	0.311	8	8	0.961
SD	41.43	29.82		26.83	14.62		7.15	7		18.94	19.24	
Range	255	200		132	67		41	32		123	134	

DS = Down syndrome; GW = general ward; ICU = intensive care unit; SD = standard deviation.

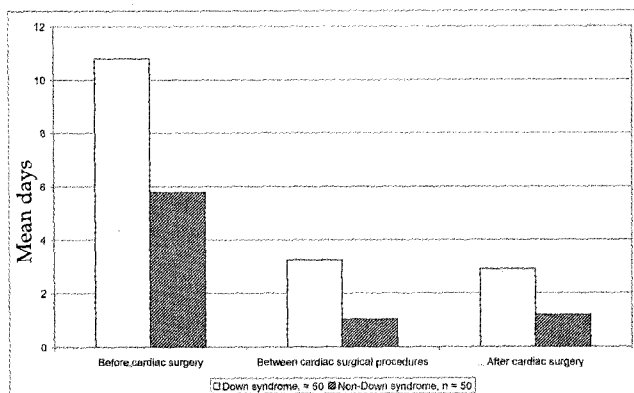


Fig. 3. Mean number of days in hospital in cohorts of Down syndrome and non-Down syndrome children who underwent cardiac surgery at Red Cross War Memorial Children's Hospital between January 1998 and June 2003.

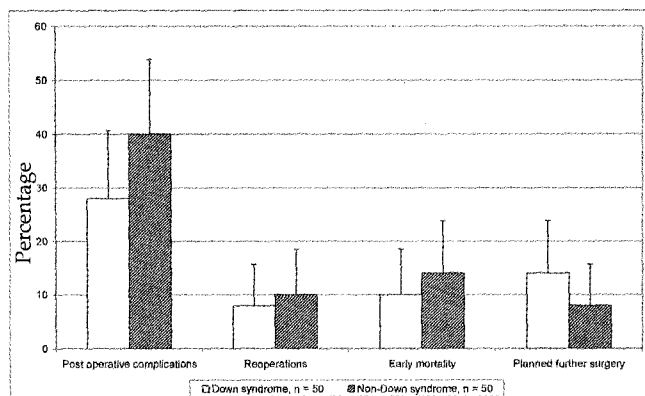


Fig. 4. Postoperative course in cohorts of Down syndrome and non-Down syndrome children who underwent cardiac surgery at Red Cross War Memorial Children's Hospital between January 1998 and June 2003. Error bars represent 2 standard deviations.

life expectancy for someone with Down syndrome is still approximately 20 years less than that of a person without Down syndrome.<sup>9</sup> This improvement has largely been attributed to advancements in cardiac surgery and to the higher proportion of institutions that are offering the same management strategy for CHD to Down syndrome children

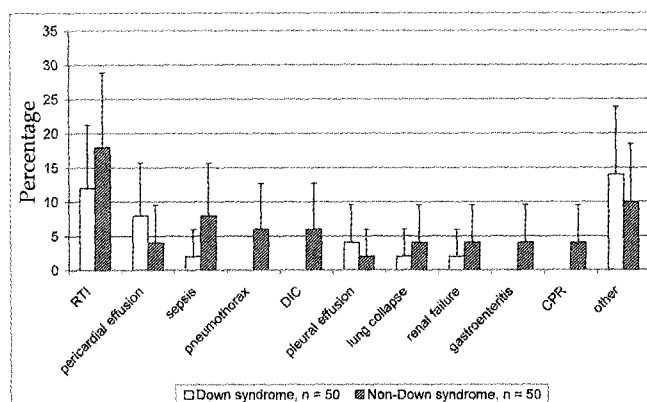


Fig. 5. Post-operative complications in cohorts of Down syndrome and non-Down syndrome children who underwent cardiac surgery at Red Cross War Memorial Children's Hospital between January 1998 and June 2003. RTI – respiratory tract infection; DIC – disseminated intravascular coagulopathy; CPR – cardiopulmonary resuscitation. Error bars represent 2 standard deviations.

as they offer to non-Down syndrome children.<sup>8,9</sup> More importantly, the survival profile of Down syndrome children with surgically treated CHD has been shown to be similar for Down syndrome children without CHD.<sup>4,11</sup> This is an indication that surgical intervention is being utilised effectively abroad.

The disparity in the treatment strategies offered by different public health institutions in South Africa may be the result of misconceptions regarding the benefit of surgical treatment of CHD of children with Down syndrome. This is evident in the statements issued by the Gauteng Provincial Government in response to the case of Baby Ronnie: that Down syndrome children represent about 30% of children with cardiac problems and that Down syndrome children have more complex heart lesions and are usually more prone to life-threatening complications as a result of surgery.<sup>12</sup>

Firstly, the prevalence of Down syndrome in all patients requiring cardiac surgery has convincingly been reported as approximately 9%,<sup>3</sup> not 30%.

Secondly, the policy of offering surgical treatment to Down syndrome children for CHD at RXH is qualified ethically

Table III. Benefit of surgery in terms of number of days spent in hospital

	Before cardiac surgery (days)			Between cardiac surgical procedures (days)			After cardiac surgery (days)		
	DS	Non-DS	p	DS	Non-DS	p	DS	Non-DS	p
Mean	10.8	5.78		3.24	1.04		2.9	1.2	
Median	2.5	0	0.002	0	0	0.846	0	0	0.172
SD	16	15.93		8.6	10.41		5.87	3.06	
Range	65	86		44	21		28	15	

DS = Down syndrome; SD = standard deviation.



by Lawrenson *et al.*<sup>6</sup> and has been qualified in terms of cost efficiency by this study and previous studies. Ungerleider *et al.*<sup>13</sup> used a cost tracking system to demonstrate that Down syndrome children had a less predictable cost picture in the repair of VSDs than children without Down syndrome, but the presence of Down syndrome did not influence the risk of higher costs in the repair of other CHDs. Our study demonstrates that Down syndrome children undergoing surgical treatment of CHD do not spend significantly more time in hospital or in the ICU than their non-Down syndrome counterparts, and thus, the two groups appear to place a similar burden on the health care system (Table I).

Weintraub *et al.*<sup>14</sup> and Rizolli *et al.*<sup>15</sup> demonstrated that Down syndrome children required fewer re-operations and compared favourably on clinical and echocardiographic investigation than children without Down syndrome in the surgical management of AVSD. In this study, the two groups showed a similar postoperative course in terms of postoperative complications, re-operations and early mortality (Fig. 4). It is important to note that the early mortality rates of 10% in the Down syndrome group and 14% in the non-Down syndrome group recorded in this study were higher than the early mortality rates recorded over the past 10 years for children undergoing cardiac surgery at RXH – the lowest annual early mortality rate was 3.6% and the highest was 9% (personal observation).

Evaluating the long-term survival of the two groups was outside the scope of this study. The expected 1-year survival rate of children with Down syndrome has been reported as approximately 90% with a 10-year survival rate of about 80%.<sup>8,9</sup> Various studies have reported a life expectancy for children with Down syndrome in the 50s<sup>9</sup> with 25 - 45% surviving to the age of 60 years.<sup>15,16</sup> It has been demonstrated that these survival rates are significantly less than those of the general population where a 60-year survival rate of approximately 85% has been demonstrated.<sup>17</sup> Recent long-term survival rates for Down syndrome people in South Africa are not available.

There are various factors that appear to be more important predictors of the outcome of cardiac surgery than the presence of Down syndrome. These include the preoperative medical condition of the child,<sup>17,18</sup> the anatomical complexity of the cardiac lesion,<sup>17,19</sup> and the timing of surgical intervention.<sup>15,18,20</sup>

The management of the preoperative medical condition is necessary for any child undergoing cardiac surgery since the high frequency of associated non-cardiac diagnoses is not unique to Down syndrome children,<sup>17</sup> and is confirmed in this study (Fig. 2). It is generally accepted that AVSDs (a complex cardiac lesion) constitute 40 - 60% of cardiac lesions in Down syndrome children with CHD while AVSDs are uncommon in children without Down syndrome.<sup>2,21</sup> However, the *most complex* forms of this lesion, together with other complex lesions such as TGA, are usually found in non-Down

syndrome children, and are less favourable to repair.<sup>2,15,19</sup> This distribution of cardiac diagnoses is consistent with the findings in this study (Fig. 1). It is well established that early repair is recommended in any child with CHD.<sup>15,18</sup> This appears to be especially important in Down syndrome children in order to prevent associated pulmonary diseases since they have a higher propensity for early pulmonary hypertension than non-Down syndrome children.<sup>3</sup>

In this study the Down syndrome group appeared to benefit more from the cardiac surgery than the non-Down syndrome group. This is evident in the finding that the Down syndrome group showed a significant reduction in the time spent in hospital after having received cardiac surgery whereas the non-Down syndrome group did not show a significant reduction. This is further evidence to support the need for cardiac surgery in Down syndrome children with CHD and may indicate that *not* operating on these children may result in more hospital resources being consumed.

The significance of the preponderance of females in both groups for this study is questionable, given the size of the study population. However, previous studies have reported similar findings.<sup>9,11</sup>

The difference in the distribution of cardiac diagnoses prevented a comparison between the two groups according to the type of defect present and type of surgical procedure required. However, as previously mentioned, the type of cardiac lesion is not one of the criteria for consideration for cardiac surgery.

Conducting a study of this nature in collaboration with other health care facilities in South Africa that offer surgical repair of CHD would provide an understanding of the problem in the broader context of South Africa.

## Conclusion

This study demonstrates that cardiac surgery in Down syndrome children does not place a significantly greater burden on the health care system than in non-Down syndrome children with CHD, and that, in this comparison, Down syndrome children appear to benefit more. The postoperative course for the two groups of children is similar. Denying cardiac surgery to children with Down syndrome may therefore not improve the efficiency of resource allocation, especially since the number of cardiac surgical procedures performed on Down syndrome children is a small proportion of the total surgical load. It therefore appears unreasonable to suggest that discriminating against children with Down syndrome will ameliorate the problem of scarce resources. For these reasons we conclude that cardiac surgery is warranted in children with Down syndrome. This review, however, does not attempt to quantify the broader health economic issues of an interventional versus a conservative approach to CHD in children with Down syndrome. This review is the first South



African audit of an interventional management programme for CHD in Down syndrome children, and falsifies the widely held misconception that cardiac surgery in Down syndrome children is more complex and costly than on non-syndromic children. This argument can therefore no longer be used for the motivation or defence of non-intervention in the South African context.

It is clear that the health care system is limited less by the technology and skills involved in treatment strategies and more by the resources available to fund such strategies as medical advancement continues with alacrity. As a result, doctors are frequently faced with moral choices in which medical needs, ethical issues and economic constraints are conflated to the degree that correct and incorrect practice and the pursuit of optimal health care become an abyss of uncertainty.

The results of this study are evidence, relevant in the South African context, of the benefit to be derived by the surgical management of CHD in children with Down syndrome, a benefit readily realised in the formulation and implementation of more appropriate policy.

'Happiness is not an ideal of reason but of imagination.'

– Immanuel Kant (1724 - 1804). *Fundamental Principles of the Metaphysics of Ethics* (1785)

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Accepted 29 June 2006.

