ETHICS

Why are some South African children with Down syndrome not being offered cardiac surgery?

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About 1 in 1 000 children has Down syndrome. Extra chromosomal material results in a myriad of potential problems for the affected individual. About 40% of Down syndrome children will have cardiac abnormalities, ranging from the simple arterial duct to the complex atrioventricular septal defect. Virtually all these defects are amenable to surgical correction and extended survival is possible. In South Africa many of these children do not undergo cardiac surgery.

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In all provinces in South Africa specialist medical services are under threat. Many of these services are perceived as being expensive and serving patients with limited outcomes. Resource constraints (in particular nursing shortages and budgetary limitations) have forced specialists to examine their practices and curtail those practices that offer low yield at high financial cost. The recently published 'head injury' protocols are a good example of such a relook.[1] Denying ventilatory support to premature infants weighing less than 900 g is another example of a policy designed to reassign resources to those who might achieve a better outcome.

All cardiac surgical units within the State sector in South Africa are affected by these resource constraints. The long waiting lists for surgery are the most obvious manifestation of these limitations. For children with congenital heart disease, a longer waiting period may mean the difference between low-risk and high-risk (and more expensive) surgery. At worst, the wait may make safe surgery impossible.

Prioritising patients within waiting lists is not an easy task. We have some experience with at least three approaches to shortening lists:

1. Increasing resources (even for a short period of time, as happened during 2000 in the Western Cape) helps to shorten the list. 'Fairness' can be achieved within a hospital setting by rotation of priorities – cardiac patients this year, tonsillectomy patients next year and so on.

2. The use of more expensive devices may allow non-surgical treatment of simpler lesions in the catheterisation laboratories rather than in the operating theatre. (While the cost of closing an atrial septal defect with a closure device may exceed the cost of the same procedure surgically in the setting of a developing country,[2] the opening of an operating space for a child with tetralogy of Fallot may offset that extra cost, in terms of future hospitalisations and morbidity).

3. Careful reorganisation of the list, which involves a thorough re-analysis of the indications for surgery and postulated outcome for each patient. It may require the wisdom of Solomon to deny surgery to patients who are perceived as benefiting less from surgery than others.

With our profession's basic commitment to preserve life for all who seek our help, selecting any patients who will not be helped is an ethical minefield. The only criterion that stands even a chance of being a 'fair' or 'just' basis for selection is the likelihood of long-term medical benefit from the care provided. There is no ethical ground for choosing on the grounds of long-term contribution to society or any other assessment of 'worth'. But how do we assess the likelihood of long-term medical benefit? Personal moral judgements (of social worth and promise) inevitably cloud assessments of medical benefit. This seems at least in part to be the problem with certain patients being refused cardiac surgery.

The 'Baby Ronnie' episode brought the (sincere) attempts of one province to shorten the waiting list of children waiting for cardiac surgery into the limelight. Baby Ronnie was flown at great cost to a private facility in the Cape when he was denied cardiac surgery in the State sector in another province.[3]

What made Baby Ronnie different from any other child with the same congenital heart defect? He was born with Down syndrome.

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The scope of the problem of children with Down syndrome and congenital heart defects

About 1 in 1 000 children has Down syndrome (most commonly due to trisomy 21).[4] The extra chromosomal material results in a myriad of potential problems for the affected individual. About 40% of Down syndrome children will have cardiac abnormalities ranging from the simple arterial duct to the atrioventricular septal defect.[5] Virtually all defects are amenable to surgical correction and extended survival is possible.

The majority of children with Down syndrome and congenital heart defects (60%) will have simpler defects such as ventricular septal defects, tetralogy of Fallot or patent arterial ducts. In our opinion there are no medical reasons that make Down syndrome patients with these particular defects less likely to benefit from surgery than their counterparts with a normal karyotype.

The other 40% of patients with Down syndrome and a heart abnormality will have an atrioventricular septal defect, which is a far more complex lesion. This abnormality in essence involves the presence of a large gap in the walls between both atria and ventricles and abnormal atrioventricular valves. (These patients have a common atrioventricular valve instead of true 'mitral' and 'tricuspid' valves.[6]) Repair of the atrioventricular defect is a significant challenge for the surgeon.

If repair of atrioventricular septal defects is delayed beyond the first year of life, irreversible pulmonary vascular changes may ensue as a consequence of the high-pressure left to right shunt. Surgery may then no longer be possible.[7] After the first year of life, if they survive congestive cardiac failure and repeated chest infections, unoperated children enter a 'honeymoon period' as they develop irreversible pulmonary hypertension. They then succumb in their third decade from right heart failure or the consequences of persistent cyanosis.

Patients with Down syndrome also have other associated abnormalities such as mid-face hypoplasia and airway obstruction that increase their propensity to develop accelerated pulmonary hypertension.[8] For these reasons, there is a perception that cardiac surgery in these patients is more hazardous and associated with a higher morbidity and mortality. This perception is incorrect.

Reasons given for not operating on Down syndrome children with an atrioventricular septal defect

Poor surgical results

In an article published in the early 1980s Bull and colleagues[9] argued that not offering surgery to Down syndrome patients with an atrioventricular septal defect resulted in equivalent survival to the surgical option. Despite cogent arguments, their ideas were highly controversial at the time. The article was published in *The Lancet* after having been rejected by the *New England Journal of Medicine*.[10]

Improvements in surgical outcome have resulted in their conclusions becoming obsolete. The life expectancy of patients with Down syndrome has doubled since the 1970s largely as a consequence of treatment of these heart lesions.[11] Most USA and UK centres would claim 30-day mortality figures of less than 5% for correction of an atrioventricular septal defect in a patient with Down syndrome.[12]

There is no reason why South African surgical centres cannot achieve equivalent surgical results (Table I). At Red Cross Children's Hospital, the mortality for isolated complete atrioventricular canal repair was 6.9% (4 of 58, of whom 44 had Down syndrome).[13]

Limited lifespan and early dementia of patients with Down syndrome

The lifespan of a person with Down syndrome is less than that of a person with a normal karyotype.[11] Nevertheless, the patient may reach an advanced age. In societies with resources, 13% of patients with Down syndrome reach the age of 60 years.[14]

Intellectual handicap is universal in individuals with Down syndrome and most children will face institutionalisation as adults. Up to 50% of patients with Down syndrome will show

Table I. Five years' experience at Red Cross Children's Hospital, comparing selected procedures with Stark *et al.

	Red Cross: 1 unit, 5 years		UK: 5 units, 1 year	
Operation	Number	Mortality (%)	Number	Mortality (%)
All operations	1 334	5.1	1 378	4.0
Isolated VSD	162	1.2	168	0.6
Fallot's tetralogy	115	3.5	88	2.3
Switch (simple TGA)	15	6.7	67	0
Complete AVSD	58	7	55	3.6

*Reproduced from: Hewitson *et al*.[13]
VSD = ventricular septal defect; TGA = transposition of the great arteries; AVSD = atrioventricular septal defect.

signs of Alzheimer's disease by the age of 50.[15] Recent data suggest that the diagnosis of dementia in these individuals is difficult and late-onset hypothyroidism as well as depression as a consequence of the death of a caregiver may cause a 'pseudodementia'.[16]

For some, sheltered employment will be possible. For all affected individuals, however, the intangible elements that make up 'quality of life' will nevertheless be present in abundance.[17]

Does not fixing a cardiac defect save money?

The patient with a significant defect is likely to suffer from cardiac failure and repeated chest infections. The 'honeymoon' period alluded to previously may take a year to develop with symptoms and signs of congestion gradually decreasing unless the patient succumbs prior to this time. Each episode of pneumonia might result in a hospital admission. Each illness may be associated with time away from work for the child's parents. The cost of an admission to hospital may rapidly start to offset money saved by not repairing a defect (see article by Roussot *et al*.[18] in this edition of the journal).

The patient with severe pulmonary hypertension is said to be relatively free from symptoms until the 2nd or 3rd decade.[9] The end of their lives is associated with worsening dyspnoea and cyanosis and life-threatening dysrhythmias; in our experience, the last few years of these adolescent children's lives are often miserable.

How many Down syndrome patients are there?

Only 8% of patients with congenital heart disease have Down syndrome (i.e. on the average surgical waiting list only 8 of every 100 patients should have Down syndrome). Therefore not performing surgery on these patients will represent only an 8% decrease in waiting lists. (If simpler lesions are corrected and atrioventricular septal defects are not, a 'saving' of 3 - 4% on a list will be achieved.)

What are the rights of patients with Down syndrome?

The rights to basic health care are guaranteed to all South Africans as a constitutional right.[19] The definition of what constitutes 'basic health care' is open to interpretation. In South African law, the Soobramoney case is cited as a legal definition of basic health care and has been used to justify withholding super specialist services.[20]

It could be argued that an expensive operation that 'guarantees' a long survival is also effective and basic health care when the costs are defrayed over a long time period. For example, the chances of surviving for more than 30 years after repair of tetralogy of Fallot are excellent.[21]

The challenge to health care planners is not only to make sure that no one succumbs from a simple pneumonia but also to ensure that the patient with congenital heart disease from Mafikeng has a similar chance to the patient from Cape Town to receive the surgery that will extend his/her life. The patients who benefit from so-called 'ivory tower' medicine do not come from a different country to the patient who dies when gastroenteritis is not treated properly.

Is it unethical not to offer surgery to South African patients with Down syndrome?

Distribution of medical care on the basis of lifespan, quality of life, or contribution to society is a slippery slope towards denying care to many other patients, such as severe burns or oncology patients to mention only two. Why should Down syndrome be singled out?

In an elegant article in the *British Medical Journal*, Savulescu[22] argues that medical practitioners are used to rationing resources. Furthermore, limiting available treatments with some benefit in favour of treatments benefiting more people to a greater degree may not be illegal despite being unethical, provided decisions taken to limit medical care are taken in consensus and that the process is transparent. In addition, the decision to limit access to a particular resource should be considered for reversal should circumstances change.

The reasons for not offering surgery to children with Down syndrome and congenital heart defects – never working, intellectual impairment, and the 'stealing' of surgery from potentially productive individuals – need to be carefully examined. In a country with 30% unemployment, 'not being employable in the open market' is a somewhat soft reason to deny surgery. In addition, the right of mentally handicapped individuals to work is protected under basic labour laws![23]

When we reject a child's candidacy for surgery because he has a low IQ, then we have allowed judgements of social worth and promise to masquerade as questions of the likelihood of long-term medical benefits.

It is disconcerting that within major urban centres there are units in the public sector hospitals which feel that they are not able to offer surgery to patients with atrioventricular septal defects and Down syndrome existing cheek-by-jowl with centres in the private sector where such children are offered surgery because they have parents with sufficient financial resources.

Conclusion

The history of the South African medical establishment is tainted by less than perfect decisions taken for political and/or financial expediency. The decision to restrict a service or withhold a particular service must be taken against a backdrop

of idealism and striving to obtain the maximum good rather than as a reaction to political or economic pressure. In the 'supermarket' of medicine, super-specialists are the 'loss leaders'. They enhance the quality of the entire system and this benefits not the health economist but the patient.

Given scarcity of resources and in the face of the need to make tragic choices about how to deal with long waiting lists, what can we do? The first thing is to be truthful: acknowledge the limits and scarcities we face to both our patients and the public. There is a reticence to speak openly in the media about the health services in crisis because it puts health politicians in a bad light. But public money is being spent on a public service about which we must be transparent. Such transparency implies a readiness to admit that there are limits to what we can do, but also that we will not turn our backs on basic patient rights.

The problems that beset South African cardiac surgery (which can be summarised as 'too few operations for too many patients with too few nurses') may in our opinion be better dealt with by reviewing our practices as a whole rather than denying surgery to one group of children with an easily identifiable genetic abnormality.

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