Paediatric cardiac surgery for a continent – The experience of the Walter Sisulu Paediatric Cardiac Centre for Africa

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ABSTRACT

Very few African countries have the resources to provide optimum paediatric cardiac services to their largely indigent populations. In the current era, in countries with access to modern paediatric cardiac care, mortality for congenital heart disease occurs more often in adulthood than in childhood. This level of care is largely unavailable in Africa. The Walter Sisulu Paediatric Cardiac Centre for Africa was set up in 2003 as a public-private collaborative initiative to extend modern paediatric cardiac care to the continent. Three core functions form the basis of our operations: service delivery, training, and research.

This communication reviews our experience with this effort over an eight-year period. We have performed 2 023 procedures on 1 738 patients including a large proportion of neonates and infants with an overall mortality of 7.1%. Our charity arm sponsored 21.5% of these patients. We have encountered problems peculiar to the African context which we discuss. We also describe innovative techniques in management of specific patient populations. Our training efforts yielded two qualified paediatric cardiac surgeons who now work at the centre and two additional surgeons are in training. We have participated in research leading to publication of papers in peer-reviewed journals. In spite of our achievements, we recognise the enormous challenges faced by the continent in terms of paediatric cardiac care. An attempt has been made to quantify the burden of congenital disease in Africa to guide planning and training. We offer recommendations on how to address some of these pressing health issues for children of the continent.

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INTRODUCTION

Congenital heart disease (CHD) is the most common congenital birth defect and an important cause of infant mortality.[1] Globally, approximately 7% of all neonatal deaths are attributable to major congenital malformations, of which at least 25% are due to severe forms of congenital heart disease.[1] In the pre-surgical era, less than 20% of children born with CHD survived into adulthood.[2] The application of the advances made in paediatric cardiology and cardiac surgery in the last 50 years now ensures that survival into adulthood occurs in 70 to 80% of children born with cardiac disease with access to available optimum treatment.[2] This level of care for CHD is unavailable in Africa.

Management of CHD often requires relatively sophisticated diagnostic techniques and high-level infrastructure operated by personnel with advanced training and expertise. The manpower and infrastructure required is largely unavailable on the African continent. While in North America and Europe there is approximately one congenital heart surgeon for 3.5 million individuals, in Africa the corresponding figure is one congenital heart surgeon for 38 million individuals, making Africa the most underserved continent in terms of congenital heart surgery.[3] To complicate the issue further, the treatment costs related to diagnosis and treatment of CHD is way beyond the means of the largely indigent population. In parts of western Africa, only 20% of the parents of children (less than 15 years old) requiring surgery for congenital cardiac disease are able to finance the operation within 12 months of diagnosis.[4]

Very few African countries have the resources to provide optimum paediatric cardiac care and large numbers of families with CHD lack the resources to access the available treatment. South Africa and Egypt have the best paediatric cardiac service on the continent and yet even in these countries, deficiencies exist. In South Africa, where probably the most comprehensive paediatric cardiac programme on the continent is housed, less than 40% of the operations required for CHD are carried out and the infrastructure and resources to detect and manage heart disease in children, particularly within the public sector, are grossly inadequate.[5]

For the rest of the continent, paediatric cardiac care is rather scant. In western Africa, Nigeria – the most populous country – performed 102 cardiac operations between 1974 and 2000.[6] Ghana performed its first open heart procedure, an ASD closure, in 1962 but its cardiac programme faltered until 1989. Since then, its National Cardiothoracic Centre, the only tertiary cardiothoracic facility in the country has been performing an increasing number of open-heart procedures. The centre currently performs about 400 cardiothoracic surgical operations annually; a quarter of these are cardiac procedures and roughly 75% of cardiac operations are for congenital heart disease and rheumatic heart disease. Civil war has destroyed much of what Cote d'Ivoire had of an active cardiac programme (personal communication F.E.). Paediatric cardiac surgery is also performed by centres in Kenya, Sudan, and Mozambique. Altogether, about 2 500 to 3 000 children undergo cardiac surgery annually on the African continent[7] (population of over one billion). With such a deficiency in comprehensive paediatric cardiac care on the continent, there is a clear need for the more resourced countries to reach out to the continent's vast numbers of children with no access or funding for paediatric cardiac care.

THE WALTER SISULU PAEDIATRIC CARDIAC CENTRE FOR AFRICA (WSPCCA)

The problem of a lack of corrective paediatric cardiac surgery on the African continent created the impetus to establish the Netcare Sunninghill Hospital Paediatric Cardiac Centre for Africa in January 2003, with three goals: firstly, the creation of a surgical hub to treat children with correctable heart disease on the African Continent; secondly, mindful of the impossibility of servicing the entire continent, to offer training and skills development to doctors on the African Continent; and, thirdly, to participate in clinical research in the African context. The institution was set up under the leadership of the senior author (RHK).

Walter Sisulu was an icon in the apartheid struggle and mentor to Nelson Mandela. With Sisulu's passing in 2003, Mandela considered that a true living memory – not a statue – was appropriate for him. Consequently, the Netcare Sunninghill Hospital Paediatric Cardiac Centre for Africa was renamed the Walter Sisulu Paediatric Cardiac Centre for Africa (WSPCCA) with the idea that neonates, infants and children from the entire continent, having had congenital heart defects corrected at the WSPCCA, would return to their native countries and "live the memory" of Walter Sisulu. Its national patron Nelson Mandela officially opened the unit on 7 November 2003, while her Royal Highness Princess Haya Al Hussein, a major benefactor, subsequently became the global patron.

While Sunninghill Hospital is the headquarters of the WSPCCA, the vision to make the facility national has resulted in operations being performed at the Christian Barnard Memorial and the Red Cross Children's War Memorial Hospitals in Cape Town, supported by the Walter Sisulu Paediatric Cardiac Foundation (WSPCF).

Each hospital has a first world unit, operating predominantly in a third world environment. At present approximately 85% of patients are self-funded (medical insurance or personal) and 15% are supported by the WSPCF through contributions from donors. In addition, in each hospital there is a "rob Peter to pay Paul system" whereby donor-funded patients intermingle imperceptibly with other patients, without any discrimination while using the same facilities.

FULFILLING THE MISSION – ACHIEVEMENTS

Service

Since its inception in 2003, our programme has performed 2 023 procedures on 1 738 patients including a large proportion (53%) of

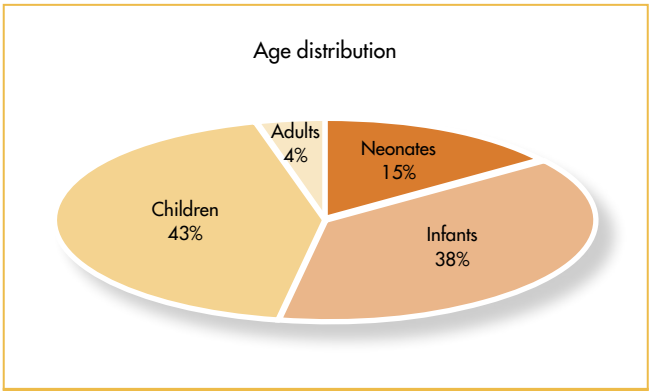

FIGURE 1: Age distribution of patients. Neonates (less than 1 month old), infants (1 to 12 months old), children (1 to 18 years old), adults (more than 18 years old).

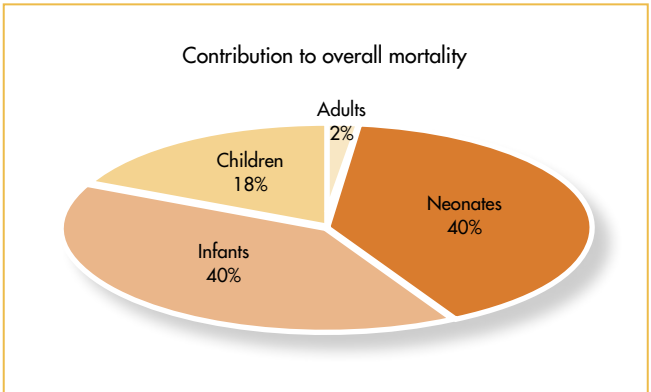

FIGURE 2: Mortality distribution for the various age groups.

neonates and infants under a year old (Figure 1) with an overall early mortality of 7.1%. Our outcomes are influenced by the relatively high numbers of neonates and infants, who together account for 80% of the overall mortality (Figure 2), a significant number of them presenting with co-morbidities such as nutritional problems, low birth weight, pre-operative infection, and HIV infection.

The caseload has grown to over 350 cases per year with a complex mix of congenital and acquired cardiac lesions (Figure 3) presenting to the centre for diagnosis and treatment. Of these patients, our charity arm, the WSPCF funded the care of 373 (21.5%) indigent patients extracted from countries across the African continent (Figure 4).

Management policy

For correctable lesions, we favour early complete correction of CHD rather than palliation. Peculiar issues have however evolved. In the context of our socio-economic background, the spectrum of cases seen and the crucial management problems that arise differ from the situation in Euro-American centres. These are important factors that affect outcomes. Late presentation is an important issue that becomes critical in lesions requiring early correction such as transposition with intact septum (TGA-IVS), truncus arteriosus, atrioventricular canal defect, and others. In truncus arteriosus for example, 45% of our patients presented beyond 3 months of age and surgical correction was achieved with an early mortality rate of 18.4%. Similarly, in TGA, the early mortality for all anatomic types was 12.5% over the entire period, a figure that has decreased markedly over the last 4 years.[8]

Technical innovations

In dealing with the issue of late presentation, we have had to modify our management approach. This is especially pertinent to the infant with TGA-IVS. With the advances made in paediatric mechanical circulatory support, we modified our policy on the treatment of TGA-IVS from August 2006. We adopted a policy of extending the age limit of the primary arterial switch operation up to 10 weeks of age, with mechanical circulatory support as a stand-by rescue. We have reported[8] results for the primary arterial switch for TGA-IVS in patients 3 to 10 weeks of age comparable to those younger than 3 weeks of age.

We have developed a method of optimal exposure of the subaortic region for the transaortic approach to lesions such as discrete subaortic stenosis, subaortic VSD, and hypertrophic obstructive cardiomyopathy. The hexagonal six-point traction technique[9] facilitates exposure of the subaortic region with minimal use of metallic retractors. Our results with this technique in the last 39 consecutive procedures evaluated have been good: early mortality of 2.6% (1 death) with a mean cross-clamp time of 38.4±16.4 minutes.

We performed a total of nine Mustard operations in the period primarily for late presenting transpositions. By extending our age limit for the primary arterial switch operation to the first ten weeks

of life,[8] Mustard operations have been performed less frequently since August 2006.

With the advances made in paediatric mechanical circulatory support, we have operated on all children referred with anomalous left coronary artery from the pulmonary artery (ALCAPA) regardless of the left ventricular (LV) function. In reconstructing the main pulmonary artery, we employ glutaraldehyde-treated autologous pericardium for pulmonary root augmentation to avoid tension on the anastomosis, stretching, and compression of the relocated coronary artery. We have obtained excellent results with this approach with a left ventricular assist device (LVAD) on standby to rescue poor LVs (lowest preoperative fractional shortening of 6%). The postoperative salvage rate using LVAD for post-cardiotomy LV failure in this patient population has been 85.7% (6 out of 7) in our experience.

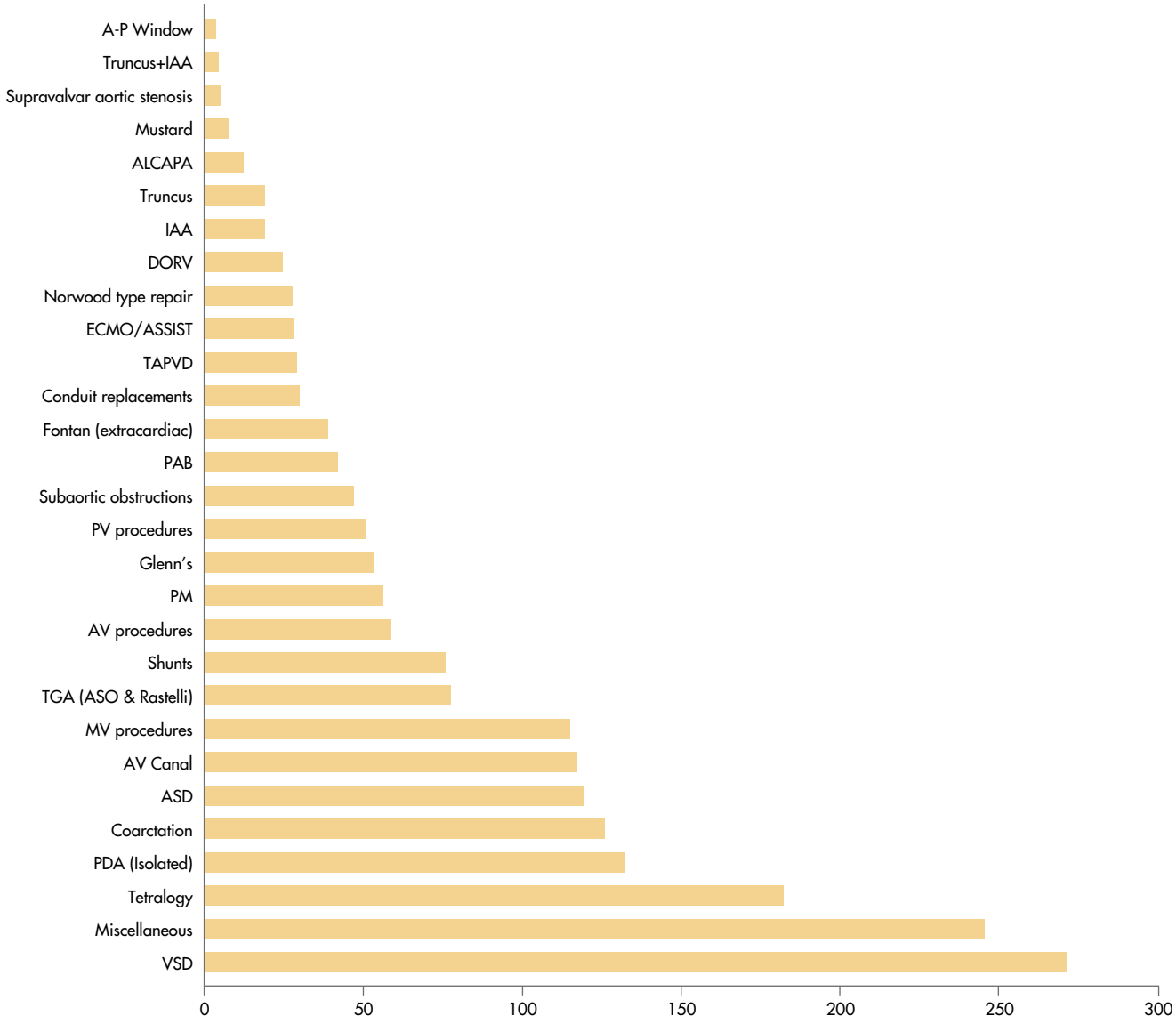

FIGURE 3: Specific procedures performed.

A-P: aortopulmonary, IAA: interrupted aortic arch, ALCAPA: anomalous left coronary artery from the pulmonary artery, DORV: double outlet right ventricle, ECMO: extracorporeal membrane oxygenation, TAPVD: total anomalous pulmonary venous drainage, PAB: pulmonary artery band, PV: pulmonary valve, PM: pacemakers, AV procedures: aortic valve procedures, TGA: transposition of great arteries, ASO: arterial switch operation, MV: mitral valve, AV Canal: atrioventricular canal, ASD: atrial septal defect, PDA: patent ductus arteriosus, VSD: ventricular septal defect.

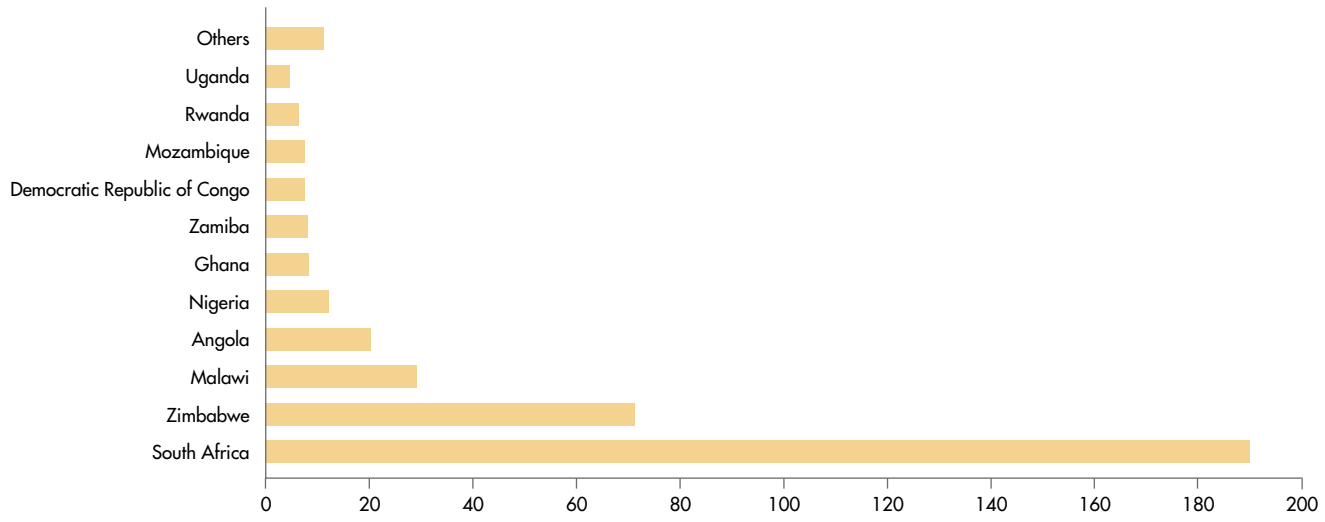

FIGURE 4: Countries served by WSPCCA from 2003-2010.

In repair of primary coarctation, we favour the use of patch aortoplasty employing oversized patches fashioned from Gore-Tex tube grafts (from 6mm for the neonate). Aneurysms have been rare in our experience and the recurrence rate over the period has been satisfactory at 6.5% (6 of the last 92 procedures) which compares favourably with a recent report.[10]

Our right ventricle to pulmonary artery (RV-PA) conduit replacement technique has evolved to our currently favoured approach of aortic transection with maximal exposure of the right ventricular outflow tract (RVOT) and pulmonary artery confluence and branches. We employ a retrograde takedown technique after which the new conduit is implanted. We elongate the aorta with a short appropriate-sized Dacron graft to relieve pressure on the implanted conduit.

Our policy for single ventricle repair currently involves early (6-9 months) bi-directional Glenn shunt and a low threshold to taking down failed Glenn and Fontan repairs. Our results for the Norwood procedure have not been satisfactory (30% hospital survival). Perhaps no lesion demonstrates the vital importance of a comprehensive approach to management of CHD better than hypoplastic left heart syndrome (HLHS). This is one area we need to improve upon in the coming years.

The availability of mechanical circulatory support for our patients has made a positive contribution to our results. This is clearly the case in the management of the child with ALCAPA where the salvage rate is in excess of 85% for patients with post-cardiotomy LV failure.

Rheumatic heart disease is still a scourge on the continent and most patients present late. There are two primary pathological processes in children with rheumatic heart disease – valvulitis and annular dilatation. All other valve pathology occur secondary to these processes. Repair is favoured whenever possible; valve replacement is reserved for patients in whom repair fails. Valve repair is tailored to the pathological findings at the time of operation. For example, a mitral valve ring is inserted to restore the shape and size of the mitral annulus. Secondary changes are treated accordingly. We believe that the prevalence of rheumatic heart disease in Africa is unacceptable and every effort must be made by governments and indeed all stakeholders to ensure that preventive guidelines are followed.

Training

We began our training programme by enlisting two locally trained surgeons in 2004, for further training in paediatric cardiac surgery. These have since become fully-fledged paediatric cardiac surgeons

on the WSPCCA team and have taken up the task of training others.

Our next two trainees joined in 2009, one from South Africa and the other from Ghana. Both are cardiothoracic surgeons seeking further training in paediatric cardiac surgery and they are close to completing a two-year programme in paediatric cardiac surgery. Typically teaching at the WSPCCA is through the efforts of highly trained staff members and consists of daily early morning ward rounds reviewing all patient data. The surgical and cardiology teams are accompanied by a paediatric pulmonologist, microbiologist, and a nutritionist. Weekly cardiology meetings with discussion of echocardiograms and angiograms, weekly surgical meetings including a monthly morbidity and mortality meeting complement the learning programme. Operative technique is taught during surgery and discussions of alternate surgical approaches are encouraged. In this manner we hope to train African surgeons for the African continent who will take their skills back to their home countries.

In 2008, we introduced our international paediatric cardiac symposium as a learning forum for paediatric cardiac surgeons and cardiologists on the continent. The symposium, which in terms of attendance has outgrown its original venue at the Sunninghill Hospital, has become a popular gathering for both local and international experts. It has attracted distinguished leaders in the field such as Christo Tchervenkov and Giovanni Stellin (2008), Richard Ohye and Christian Schreiber (2009), Richard Jonas, Christian Brizard, Christopher Knott-Craig and Christian Schreiber (2010). The proceedings of the 2010 symposium were reported in the World Journal for Paediatric and Congenital Heart Surgery.[11] We have also established a yearly skills exchange programme for our clinical personnel (surgeons, cardiologists, anaesthesiologists, perfusionists, and nurses) with centres in Germany, the UK, and the US. We continue to learn from their experiences shared in their reports on return.

Research

Research in Africa is often relegated to an extremely low priority. In the midst of the several pressing socio-economic challenges facing the continent, research receives sponsorship primarily for diseases like malnutrition, diarrhoea, tuberculosis, and HIV. Cardiovascular research in many African countries receives hardly any sponsorship. Not surprisingly, the contribution of Africa to global cardiovascular research in the number of published articles between 1995 to 2002, was estimated in one report to be 0.3%.[12] In the same study period, the report suggested that developing world regions, with the important exception of Africa, achieved an increasing rate in the number of published articles. It would appear that in Africa, we are close to fulfilling the words of Albert Starr: "… practicing surgery in an intellectual vacuum".[13] Much remains to be done in the field of cardiovascular research in Africa if we are to overcome our peculiar healthcare challenges.

We have embarked on cardiovascular research in partial fulfilment of our mission. Our research activities have led to several publications[8,9,11,14-17] in peer-reviewed journals. We continue to embark on research in other areas.

THE CHALLENGE OF PAEDIATRIC CARDIAC SURGERY IN AFRICA

We have made satisfactory progress in the three areas that constitute our core mission – service, training, and research. However, our achievements pale in the light of the enormous challenges confronting the continent in the area of paediatric cardiac services. Fundamental to our continent's problem is the fact that we must practice with very little scientific data. Rheumatic heart disease continues to plague most African nations with no sign of abatement. The incidence of rheumatic fever in Africa may be as high as 30.4 per 1 000 in schoolchildren.[18] The burden of congenital heart disease in Africa is not known and so determining the manpower and infrastructure required to adequately confront this challenge must be based on conjecture. The United Nations Department of Economic and Social Affairs[19] estimate Africa's population at end of 2010 to be in excess of 1 billion, which constituted 15% of the world's population. Despite regional variations in environmental factors and genetic makeup, the incidence of congenital heart disease around the world remains consistent at approximately 0.8% of live births.[20] Considering the fact that Africa can only boast of roughly 1% of the global pool of cardiothoracic

surgeons,[21] this is indeed a heavy burden, Hart's inverse care law[22] "the availability of good medical care tends to vary inversely with the need for it in the population served" would apply to much of Africa. It is our conviction that by providing an avenue for training of cardiovascular specialists, surgical hubs can be established in the regions of Africa where cardiovascular services are most needed but least available.

We infer from our experience that four primary obstacles must be overcome to enable progress in paediatric cardiac service delivery in Africa. These obstacles are in the area of manpower development, infrastructural layouts, healthcare financing, and staff retention programmes.

Personnel development

Personnel development involves selection and training of appropriate cardiovascular teams to deliver diagnostic and therapeutic services to their populations. Already established centres of excellence on the continent can help by providing training opportunities to personnel from deprived regions of the continent.

Healthcare infrastructure

Cardiovascular care unfortunately requires a heavy financial outlay to provide the necessary physical structures that make healthcare delivery efficient and safe. Gross national product (GNP) in Africa is less than $1 800 per inhabitant per year compared to between $24 000 to $31 000 in North America and Western Europe.[21] The worldwide distribution of cardio-thoracic surgeons very closely follows the distribution of GNP,[21] a testament to the fact that cardiothoracic surgery requires a solid infrastructural basis to practice.

In this regard, the adoption of the concept of regionalisation may be helpful. Regionalisation of cardiac care is the process of creating and maintaining a comprehensive, coordinated cardiac programme to provide care to a region or population of patients.[23] Population and birth rate data can guide the geographical location of such treatment hubs to balance optimal care and travel distances for families. Such an approach requires the combined commitment of cardiovascular specialists, health institutions, and governments.

Healthcare financing

Healthcare financing continues to be a thorny issue for most developing countries. State health insurance is non-existent in most African countries and most insurance schemes in Africa, if they exist at all, do not cover paediatric cardiac surgery. Financing paediatric cardiac surgery is thus left on the feeble shoulders of the largely indigent affected families. Occasionally philanthropic organisations, charitable individuals, public appeals for funds, and subsidies from healthcare institutions become the mode of financing the near-terminal child. Until African governments improve the economies of their countries, it appears the African child with CHD will continue to depend on the benevolence of others for care. The WSPCCA model is helpful and we recommend it, but in the long term, Africans must be economically empowered to solve their own healthcare problems.

Retention of trained staff

In the absence of adequate remuneration and infrastructural layouts, trained cardiovascular specialists will tend to drift to regions of the world where these conditions are met. The brain drain syndrome is a perennial problem and poor countries are forever losing their "brains" to rich ones. This however must not serve as an excuse for African governments not to invest in training their own staff. There are no simple solutions to the brain drain syndrome in Africa. "Push" and "pull" factors have been described[24] to characterise the dynamics of this phenomenon but it appears the gross economic and political differences between Africa and the developed world has established gradients that only facilitate Africa losing its brains.

CONCLUSIONS

The Walter Sisulu Paediatric Cardiac Centre's experience shows a promising public-private collaboration model that may be reproduced in the fashion of regionalisation to serve the African population in terms of much needed paediatric cardiac services.

CHD in Africa accounts for a sizable proportion of the global CHD burden and yet has only 1% of the world's population of cardiothoracic surgeons. Centres of excellence on the continent can help

relieve this burden by offering training opportunities to Africans in the most deprived regions of the continent.

It is feasible for African cardiovascular specialists to undertake care of the child with serious forms of heart disease on the continent with acceptable results.

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