Priorities in childhood cancer

Almost 70 delegates gathered in Stellenbosch in early September at the sixth biannual meeting of the South African Children's Cancer Study Group (SACCSG). Established in 1987, the organisation brings together paediatric oncologists, paediatric surgeons, radiation oncologists and pathologists. These meetings are an opportunity to share outcomes, discuss new treatments and enjoy the contributions of invited speakers.

When the SACCSG meets, it focuses on a group of rare diseases that cannot be prevented by lifestyle change, are almost never amenable to screening tests and tend to occur in deep-seated organs, making diagnosis difficult. However, these diseases respond well to treatment with a survival exceeding 75% in the USA.[1]

The most common childhood cancers are leukaemias and brain tumours, with the balance consisting of various forms of lymphoma, embryonal tumours of the sympathetic nervous system, kidney, muscle, liver and retina, and bone tumours. For most paediatric malignancies, treatment is multidisciplinary in nature encompassing chemotherapy, surgery and radiotherapy. The rise of major co-operative groups in Europe and North America using standardised chemotherapy protocols has been the major driving force behind the dramatic improvement in outcomes for children with malignancy over the past four decades. These protocols continue to be improved by means of randomised trials. In addition, better health infrastructure has allowed for earlier diagnosis, increasing the number of low-stage tumours, which translates into better results.

Paediatric oncologists in South Africa use a variety of internationally accepted treatment regimens and are achieving acceptable outcomes in many cases. There are, however, no national standardised protocols for the treatment of any given malignancy. Late presentation due to a low index of suspicion, both in the community and the health services, continues to limit efforts to improve outcomes for South African children with cancer.

Since the inception of the SACCSG we have witnessed a number of milestones. A children's cancer registry was established at the outset, and continues to gather vital data. A national network of paediatric oncology centres has emerged. These centres operate in academic complexes and harness multidisciplinary teams that have developed considerable expertise in treating paediatric malignancy. In addition, supporting specialties such as radiology, intensive care and cardiology have been able to develop skill in both diagnosing malignancy and treating its complications.

A variety of efforts have been and continue to be made to improve awareness of childhood cancer. A joint initiative with CHOC (the Childhood Cancer Foundation of South Africa) has brought the St Siluan signs to the attention of the public (www.choc.org.za), and the SACCSG has publicised the warning signs of childhood cancer for health professionals via the *SAMJ*[2] and its website (www.saccsg.co.za). The organisation has in recent years begun to help oversee College of Medicine examination and certification of the next generation of oncologists.

Since the first continental meeting of the International Society of Paediatric Oncology (SIOP) was held in South Africa in 1994, South Africans have become increasingly involved in efforts to improve the outcomes for young Africans with cancer. This has taken the form of treating patients from neighbouring countries, advising African colleagues and, in some cases, providing training to African paediatricians. Recently, Peter Hesseling has spearheaded an initiative to provide rational treatment for Burkitt's lymphoma in Malawi[3] and some provinces of the Cameroon, and Janet Poole will assume the presidency of SIOP Africa later this year.

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The focus of our latest meeting was to set the stage for the development of national protocols for paediatric malignancy. The National Wilms' Tumour Working Party presented the proposed Wilms' tumour protocol, and we were privileged to host one of Europe's foremost experts on Wilms' tumour, Kathy Pritchard-Jones, as a keynote speaker. One of the most heartening aspects of the 2007 meeting was that oncologists, radiotherapists and ophthalmologists came together to discuss the treatment of retinoblastoma. These discussions should form the basis of a common national protocol for this disease.

We enjoyed a large number of high-quality inputs, both from our own members and from invited speakers. Guest speakers spoke about high-risk rhabdomyosarcoma, radiotherapy for brain tumours, the role of transplantation in hepatoblastoma and the implications of DNA damage and repair for clinical oncology. There was an engaging bench to bedside session with a talk on radiological lessons in oncology and a South African perspective on solid tumour molecular pathology.

The wide diversity of talks given by both local and foreign invited speakers, as well by our own members, highlighted the broad spectrum of clinical challenges faced by South Africa's paediatric oncology community. Faced with such diversity it behoves us to identify key priorities:

1. Achieving early diagnosis

It is an ongoing necessity to continue alerting members of the public and medical staff alike to the warning signs of childhood cancer. Paediatric oncologists have a special responsibility to educate medical students and nurses about the warning signs of childhood cancer. We also have a powerful ally in the form of CHOC, an organisation dedicated to creating and maintaining public awareness.

2. Optimising reporting to the registry

One of the most limiting factors for the planning of services for childhood cancer is the considerable underascertainment of cases in South Africa. The incidence of cancer reported in Western Europe and North America varies from 130 to 150 cases per million children aged 0 to 14 per year.[4,5] According to Census 2001, just over 14 million of the almost 45 million South Africans (32%) were under the age of 15.[6] Assuming a conservative incidence rate of 120 per million, there should be about 1 700 cases annually. The number of cases reported to the SACCSG Cancer Registry averages just over 600 per year. What could explain such underascertainment? Firstly, there may be less cases of childhood cancer, as there are no accurate population-based registers in the developing

world for comparison. Secondly, there must be considerable underdiagnosis. Thirdly, there are children being treated by surgeons, radiation oncologists and adult haematologists who are not being reported to the registry. We need to get every South African under the age of 15 with malignancy onto the SACCSG registry.

3. Ensuring that children are treated by paediatric experts

All children with malignancy should be treated by a paediatric oncologist. However, geography and logistics dictate that this is not always possible. We would argue strongly that where children are being treated by adult physicians or general paediatricians, they should be treated on paediatric protocols in consultation with a paediatric oncologist.

4. Introducing national protocols

There is now a commitment to achieve national protocols for a variety of paediatric malignancies. A South African Wilms' tumour protocol is in the process of being drafted and the trial should be open for enrolment during the course of 2008. In a similar fashion SACCSG hopes to be the catalyst for the establishment of common protocols for the treatment of retinoblastoma, acute lymphoblastic leukaemia and HIV-related malignancies.

5. Treating HIV-related malignancy

This emerging group of tumours deserves special mention. With the roll-out of highly active antiretroviral therapy (HAART) we are likely to see a growing number of HIV-related malignancies. While it is well established that HAART decreases the incidence of these tumours, a large cohort of HIV-positive long-term survivors will translate into more cases of lymphoma and Kaposi's sarcoma. We are now in a unique position to gain experience in treating these children and in turn to share that knowledge with other paediatric oncologists in the developing and developed world.

Conclusion

The controversial question remains: Why spend resources on paediatric malignancy in a country where children still die of malnutrition and communicable diseases? In 1995 Richard Cohn, somewhat prophetically, cautioned that funding might be reduced for an expensive subspecialty affecting a small number of the population, despite the fact that the majority of patients are cured.[7] Maintaining the capacity to treat children with rare diseases is not a choice we can put off for a time when our health system is better resourced. Once the skills and experience are lost they are expensive to replace. We must do what is best with what we have for each of our children.

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