8. COMPREHENSIVE CARE CENTRES: CREATING A CANADIAN NETWORK

Principles

- To outline methods of improving healthcare delivery through comprehensive care centres.

Recommendations

- All patients with SCD should have access to knowledgeable, well-equipped acute care services 24 hours a day, seven days a week.
- The acute care team should have access to advice from SCD physicians and/or other SCD medical experts (e.g. specialist NPs) 24 hours a day, seven days a week.
- All patients with SCD should have access to care by a multidisciplinary team at a SCD comprehensive care centre.
- The comprehensive care team should include: specialized physician expertise (e.g. hematology and/or internal medicine or pediatrics), nursing, social work, psychological counselling, and various medical and surgical specialists (including, but not limited to nephrology, cardiology, orthopaedic and general surgery, obstetrics, and ophthalmology).
- Due to the multisystem nature of SCD, comprehensive care centres must have access to all relevant diagnostic testing and treatment modalities. These include:
  - Diagnostic testing: transcranial Doppler, MRI (including brain MRI/MRA, and validated MRI measurement of hepatic and cardiac iron overload), echocardiography, right heart catheterization, pulmonary function, and bone mineral density.
  - Therapies: RBC phenotyping and/or genotyping, with provision of antigen-matched RBC transfusions by an expert transfusion medicine team, RBC exchange transfusion, hydroxyurea, chelation therapy, penicillin prophylaxis, immunizations.

Background

Sickle cell disease is the most common genetic disease in the world and the quintessential multisystem disease: due to the effects of chronic vascular damage and hemolytic anemia, patients may develop complications in any organ. Living with a chronic and debilitating disease in turn increases the risk of psychological and social dysfunction. All told, it has been estimated that the lifetime costs of providing care to a patient with sickle cell disease approaches $9 million (1), and this figure excludes the societal costs of unemployment and decreased work productivity. As the number of patients with sickle cell disease in Canada continues to grow, the associated burden on the medical system is becoming considerable.

Better Outpatient Care = Enhanced Outcomes and Reduced Costs

However, there is also an enormous opportunity to decrease the health care costs of patients with sickle cell disease while simultaneously improving clinical outcomes and quality of life. Multiple studies have shown that patients with sickle cell disease rely heavily on acute care medical services such as emergency room visits and inpatient hospitalization.[1-12] Inpatient hospitalizations are particularly expensive: a recent study, for example, found that the median cost of a hospital admission for a child with sickle cell disease in the US is approximately $10,000 USD, with a 90% percentile cut point of approximately $32,000 USD; moreover, hospitalization costs steadily increase with patient age.[13] Shifting the care of patients with sickle cell disease from an inpatient to outpatient setting, primarily through the provision of basic preventative health maintenance and the use of hydroyxurea, can reduce total healthcare costs by 20%.[14]

Similarly, providing a specialized outpatient treatment facility for sickle cell patients experiencing painful vaso-occlusive episodes can decrease the burden on already over-crowded emergency departments and inpatient wards. The establishment of a day treatment program at the Bronx Comprehensive Sickle Cell Center, for example, resulted in 40% fewer emergency room visits, and patients seen in the day-clinic were five-times less likely

to be admitted to hospital. Moreover, the increased expertise of the day-centre clinic allowed them to provide consultative assistance to staff caring for inpatients admitted with sickle cell crises, which resulted in a 1.5 day decrease in average length of stay.[15] The scale of financial benefit following the establishment of sickle day-hospitals has since been confirmed by centres in Boston[16], Birmingham, UK[17] and Houston.[18]

SCD Comprehensive Care Centres: Outcomes from the UK and USA

Given its complexity and multisystem manifestations, the provision of preventative and specialized care for sickle cell patients necessary to allow such gains requires the establishment of comprehensive treatment centres, in which patients have ready access to a multidisciplinary care team, which should include specialized nursing expertise, social work, psychological counselling, and various medical and surgical specialists including hematology, nephrology, cardiology, orthopaedic and general surgery, obstetrics, and ophthalmology. Such was the conclusion of the United States government in 1972, when it passed the National Sickle Cell Anemia Control Act with the goal of creating comprehensive treatment centres in that country, and in 2004 when it sought to expand and provide national coordination of this network through passage of the Sickle Cell Treatment Act.[19] The need for comprehensive care has also been echoed by the American Academy of Pediatrics[20], the United Kingdom's National Health Service[21,22] and the World Health Organization.[23]

Evidence continues to accumulate that provision of comprehensive care for sickle cell disease reduces costs as well as improves clinical outcomes.[23-26] As an example, the establishment of a comprehensive sickle cell clinic in 1999 at St. Thomas' Hospital in London, England, was followed by an approximate 60% reduction in hospital admissions, with average length of stay amongst those who were still hospitalized reduced on average by nearly one week.[24] Similarly, provision of care to pediatric sickle cell patients in Texas via a Patient-Centred Medical Home (which may be considered another vehicle for the provision of comprehensive, multidisciplinary care) decreased both individual patient emergency room visits and inpatient hospitalizations by 50%.[24]

The Need for Comprehensive Care Centres and a National Strategy for SCD

Despite its clear benefits, both to patients and to the health care system as a whole, many patients with sickle cell disease continue to lack comprehensive care. A 2007 summit on sickle cell disease organized by the American Society of Pediatric Hematology/Oncology noted that "despite the substantial advances in the treatment of SCD which have occurred in the last 30 years, there is a lack of equity in the quality of clinical care provided to patients with SCD as evidenced by a failure to apply knowledge obtained from research to much of the at-risk population."[27] A similar conclusion was reached by L Smith et al in a 2006 publication in Pediatrics, in which they state "It is the minority of sickle cell patients who receive care in one of the comprehensive sickle cell centers… There is also a crucial need to increase the workforce capacity to care for adult patients with SCD to provide appropriate continuity of care for adolescents transitioning to adult care."[19] The situation, unfortunately, is no better in Canada. In 2003, for example, a study by the Toronto District Health Council confirmed that the majority of Canadian patients with sickle cell disease reside in Ontario, and of those most lived within the Greater Toronto Area. However, only 41% of hospital visits by these patients were at one of the two Toronto comprehensive treatment centres (the Hospital for Sick Children and the Toronto General Hospital), with the remaining patients scattered amongst a large number of community hospitals where management was limited to the treatment of acute care issues.[28] Twelve years later, with approximately 100 children with sickle cell disease born in Canada every year and an untold number of additional patients arriving through immigration, progress has been frustratingly slow. While the number of hospitals providing dedicated treatment to patients with sickle cell disease has increased, it is unclear how many of them meet the standard of comprehensive care and, more importantly, Canada remains without a national strategy to ensure equitable, coordinated and effective care for all patients.[29]

Following the Canadian Hemophilia Treatment Model for SCD and Other Hemoglobinopathies

In creating a national network of sickle cell disease comprehensive care facilities, Canada already has an established model to draw upon: that which has been provided since the 1970s to patients with haemophilia and other inherited bleeding disorders. Currently, there are 25 comprehensive treatment centres in place across the country for patients with these conditions, each staffed by a haematologist, nurse coordinator, physiotherapist,

psychologist or social worker, a data manager, and other specialists as required.[30] The effect over time on health care utilization has been dramatic: patient visits to the emergency department are now a rarity for patients with hemophilia. In addition, increased patient visits to the comprehensive treatment centres have been offset by increased patient self-care, with supervision provided through telephone, internet and tele-health communications. A similar program can and should be put in place for patients with sickle cell disease.

In many cases, it would be most efficient to incorporate the coordinated care of patients with sickle cell disease with that of patients with thalassemia, another inherited disorder of haemoglobin with multisystem complications (due, in this case, primarily to iron overload). A comprehensive care program for patients with hemoglobinopathies could also be expanded, depending upon local demographics, to patients with other rare blood disorders requiring comprehensive care, such as Diamond-Blackfan anemia, hereditary angioedema and primary immunodeficiency.[31] However, it is important to recognize that sickle cell disease is not, itself, a rare disorder: it has been estimated that there are over 100 million affected individuals world-wide, with another 275 000 children born with the condition every year.[32] While the prevalence in Canada continues to increase, the more important change has been in the attitudes of the medical profession and policy-makers, who increasingly recognize how much room for improvement there is in the care provided for this population. Creating a network of comprehensive treatment care facilities is an important step in closing this gap and aligning Canada with other countries, such as the United States and United Kingdom, who face similar challenges.

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