Evaluation of Outcome of Health Care in Patients with Sickle Cell Disease Registered to Pediatrics Hematology/Oncology Unit of Ain Shams University Hospitals

Thesis
Submitted for Partial Fulfillment of the Master Degree
in Pediatrics

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2010

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رسالة

توطئة للحصول على درجة الماجستير فى طب الأطفال

مقدمة من

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٢٠١٠

The Vision, Mission and Values of Faculty of Medicine, Ain Shams University

- ***Our vision is***

To be the first in the Middle East to produce doctors with a competitive edge and lead the development of medical education.

- ***Our mission is***

To prepare a graduate having competitive skills on the national and local levels, capable of teaching, learning and training for life and is committed to the standards of medical service and professional ethics.

The College also seeks continued development of programs and courses, supports and develops scientific research with the expansion of applied scientific research and health care programs to serve the needs of society and environment development.

The College also aims to provide excellent academic staff and research faculty members, to support the upgrading of administrative and institutional systems and to provide its own resources in order to achieve the goals and objectives.

- ***Our values:***

We carry out our job aiming at **excellence** and not just performance, we practice **honesty** in everything we do, we always strive to achieve **equality** of rights and the **balance** between the right and duty, with **mutual respect** and we **work together** for the benefit of one and all.

I would like to express my deepest thanks and profound gratitude to **Professor Dr. Safinaz Adel El Habashy,** Professor of pediatrics, Ain Shams University. It was such a great honor to work under her kind guidance. Her continuous help, combined with her sincere support obliged me to bear the responsibility toward this study.

I am sincerely thankful to **Professor Dr. Sahar Mohamed Sabbour,** Prof. of Community Medicine, Ain Shams University, for sharing her expertise and valuable time, for her helpful suggestions and continuous interest throughout the course of this work.

I am greatly indebted to **Dr. Nevine Gamal Andrawes,** Lecturer of Pediatrics, Ain Shams University. To her I owe much more than I could express and much less than I could repay except in part by the satisfaction of seeing this thesis come true.

I am thankful to all patients accepted to be involved in this study.

Last but not least, I would like to thank **my family** whose love, support and encouragement enabled me to accomplish this work.

بسم الله الرحمن الرحيم

قَالُوا سُبْحَانَكَ لاَ عِلْمَ لَنَا إِلاَّ مَا عَلَّمْتَنَا
إِنَّكَ أَنتَ الْعَلِيمُ الْحَكِيمُ

صدق الله العظيم

البقرة الآية ٣٢

Introduction

The goal of healthcare, for children and everyone else, is to maximize health, not simply to treat disease. For the care of children, maximizing health entails fostering growth and development, to a great extent. As a result, health supervision and promotion are central to children's health services.

Quality of care is the extent to which health services are likely to achieve their stated goal, or, as stated by the Institute of Medicine, "the degree to which health services for individuals and populations increase the probability of desired health outcomes and are consistent with current professional knowledge" **(Lohr, 1990).** Health services are processes of care that take place in the context of a structure, that is, inputs and system characteristics, and result in an outcome (or, more precisely, a change in likelihood of an outcome) **(Donabedian, 1980).** In order for health services to achieve their intended goal, they should be necessary and appropriate, of high technical quality, and delivered with dignity and respect **(Brook, 1991).**

The mechanism for monitoring and improving the quality of a system is one key administrative characteristic of the system one of the Methods for Improving Quality of Care in Health Systems is use either scientific evidence or patient-derived information to specify targets for improvement **(HSR:**

Health Services Research 33:4 (October 1998, Part II,p 1091-1096)

Sickle cell disease (SCD) is a collective term for a group of genetic disorders characterized by the predominance of hemoglobin S (Hb S). These disorders include sickle cell anemia (SS), the sickle beta thalassemia syndromes (S β+ or S β0), and hemoglobinopathies in which Hb S is present in combination with another variant hemoglobin. The most common examples include hemoglobin SC disease, hemoglobin SD disease, and hemoglobin SE disease **(Virginia department of health, November 2005)**

Sickle cell anemia [1,2,3,4] **(1) Ronald L. Nagel, Department of Medicine(Division of Hematology),Albert Einstein College of Medicine, Comprehensive Sickle Cell Center, The Bronx, NY,10461, USA published in J. Mol. Biol. (2007) 365, 425–439) 2. Stuart, M. J. & Nagel, R. L. (2004). Sickle-cell disease. Lancet, 364, 1343–1360. 3. Sergeant, G. R. (1997). Sickle-cell disease. Lancet, 350, 725–730. 4. Bunn HF. Pathogenesis and treatment of sickle cell disease. N Engl J Med 1997; 337 (11): 762-9)** is a genetic (inherited) disease caused by a single mutation from glutamate to valine at the sixth site of the hemoglobin β chain,3 resulting in pro-duction of abnormal hemoglobin, HbS.4 One of the basic events in sickle cell anemia is the polymerization of HbS inside the red blood cells which occurs upon deoxygenation of hemoglobin. This

ultimately distorts the erythrocyte membrane and creates a rigid 'sickled' red cell.

The protean clinical manifestations of SCD result from variable degrees of hemolysis and intermittent episodes of vascular occlusion that cause tissue ischemia and acute and chronic organ dysfunction. Consequences of hemolysis may include chronic anemia, jaundice, predisposition to aplastic crisis, cholelithiasis, and delayed growth and sexual maturation. Vaso-occlusion and tissue ischemia can result in acute and chronic injury to virtually every organ of the body **(Miller et al., 2000).**

Sickle cell disease requires specialized comprehensive care to achieve an optimal outcome. Comprehensive medical care includes ongoing patient and family education, periodic comprehensive evaluations and other disease-specific health maintenance services, timely and appropriate treatment of acute illness, and genetic counseling **(Lane et al., 2001).** Appropriate treatment requires the active involvement of health care professionals with expertise in the management and treatment of SCD, usually a pediatric hematologist-oncologist working in conjunction with a multidisciplinary team **(AAP, 2002).**

In addition to medical treatment, the management of sickle cell disease requires sensitivity to important psychosocial implications of the disease and services to address them. Failure to appreciate ethnic and cultural differences between providers

and patients and families may also contribute to misunderstanding and lack of trust. Thus it is imperative that providers take time to listen to the concerns of patients and families, that they be sensitive to psychosocial as well as medical needs, and that they assist families in accessing available resources as needed **(Lane et al., 2001).**

Six core outcomes for Children with Special Health Care Needs under the federal Healthy People 2010 Objectives provide guiding principles for the care of children with sickle cell disease;

1. All children with sickle cell disease will receive regular and ongoing comprehensive care within a medical home.

2. All families of children with sickle cell disease should have adequate private and/or public insurance to pay for the services they need.

3. All children with sickle cell disease will be screened early and continuously for special health care needs.

4. Services for children with sickle cell disease and their families should be organized in ways that families can use them easily.

5. Families of children with sickle cell disease will participate in health care decision-making at all levels and will be satisfied with the services they receive.

6. All youth with sickle cell disease will receive the services necessary to make appropriate transitions to all aspects of adult life, including adult health care, work, and independence. ***(*www.healthypeople *2010)***

Aim of work is to assess disease-specific measures of health care provided to SCD patients against standards of care to know gaps in service for better planning and continuous improvement.

Study population: all patients with SCD registered to pediatric Hematology/oncology Clinic of Ain Shams University Hospitals, will be enrolled in the current study.

Plan of work:

To collect data and evaluate current service, the following methods will be included;

1- Revision of patients' files for the following data;

a) Confirmation of diagnosis.

b) Initial outpatient visit, at what age, information given (any written documents)

c) Penicillin prophylaxis (or alternative in allergy); when started, and dose.

d) Immunization; pneumococcal, hemophilus influenza, and influenza.

e) Documentation of weight and height on centile charts. Bone age for those with short stature

f) Record of spleen size in patients' notes.

g) Blood pressure record annually.

h) Individual treatment and care plan.

i) Use of hydroxyurea; any included protocol.

j) Use of blood transfusion; protocol included in file.

k) Frequency of annual review; how many times per year, predetermined or emergency visit.

l) Psychological assessment. Check for those with cerebro-vascular stroke.

m) Transition to adult care. How?

2- Patients and parents interview to complete a questionnaire designed to assess current service and get data about breaking the news, and education given. This form is designed to help facilitate understanding of the family's circumstances, knowledge of sickle cell disease, and satisfaction with health care and to identify patient and family concerns and potential barriers to appropriate

treatment. It should be completed by the family when the child is not ill (e.g., in the waiting room prior to a clinic visit) and subsequently reviewed with the family by a health care provider.

Please answer the following questions by circling yes or no

- Do you have any problems getting good health care for your child? Yes No

- Do you feel comfortable with how well you can treat and control your child's pain at home? Yes No

- Do you know how to take your child's temperature? Yes No If your child is less than 5 years old, can you feel the belly for enlargement of the spleen? Yes No

- Are you comfortable with your understanding of sickle cell disease? Yes No Do you want more general information? Yes No

- Do you need more information on how sickle cell disease is inherited? Yes No

- Do you have problems with health insurance? Yes No with transportation? Yes No

- Do you feel your child's pain problems are treated well when your child is in the hospital? Yes No

- Are you comfortable with the staff's knowledge of sickle cell disease and the way they treat your child's pain? Yes No

Do you feel that the people who work at our clinic understand and are sensitive to your cultural background and needs?

Yes No

Do you feel that you have the opportunity to take part in making decisions about your child's health care? Yes No

Would you like more contact with another family who has a child with sickle cell disease? Yes No

What is your child's grade in school? _____

Do you feel there is a need for a better understanding of your child's special needs at school? Yes No

About how many days did your child miss from school last year? _____

If your child is more than 12 years old, are you receiving services to help your child prepare for an independent adult life? Yes No

Are your other children having any problems because of their brother or sister with sickle cell disease? Yes No Are there any other worries in your life? Yes No

Would you be willing to work toward getting better care and more research on sickle cell disease? Yes No

What are the hardest things about sickle cell disease that you have to deal with?

_____

_____

_____

_____

What else can we do for you?

_____

_____

Name of child: _____

Age: _____ Date of Birth:_____

Who completed this form? (Name, relationship to patient)

Date: _____

3- Meeting care givers; to fill data about facilities available, obstacles to deliver care, local guidelines and procedures available for investigation and management of chronic problems of SCD namely; growth delay, nocturnal enuresis, renal problem, stroke, lung disease, biliary disease, avascular necrosis, and what policy they adopt for those who do not regularly attend clinic.