SICKLE CELL DISEASE
IN THE ERA OF NATIONAL TRANSFORMATION

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INTRODUCTION

- Sickle cell is global disease of old world and immigrants to the new world.
- Sickle cell anemia to predict that the number of cases in newborns is likely to rise to about 404,200 in 2050, up from 305,800 in 2010\(^{(1)}\)
- It has linear relation with previously endemic with Malaria as protective mutation to preserve the native population.

1. T Biswas. Global burden of sickle cell anaemia is set to rise by a third by 2050. BMJ 2013;347:f4676
Sickle cell gene distribution and Malaria
S haplotype background (BitounGUI et al, 2015)
INTRODUCTION

- Sickle cell disease management is a comprehensive, multidisciplinary approach which includes a collaborative effort between patients, family members and the healthcare team.

- The management is not only physical, Psychologic aspect is high.

- It need dedicated team for proper management.

- Despites efforts in research and funds, many patients do not receive proper medical care.
INTRODUCTION

- The American family medicine association:
- Primary care physicians should be familiar with SCD:
  - Pathophysiology
  - Diagnosis, evaluation.
- Current standard of care (basic care, vaccinations, periodic investigations).
- Medical management of sickle cell disease.
- When to refer.\(^{(1)}\)

2. ASH Education Book December 6, 2013 vol. 2013 no. 1 433-438
INTRODUCTION

- Unfortunately primary care physicians and workers has less knowledge about the health care delivery for SCD patients.

- Efforts made to transform primary care into a centerpiece for improving health care quality in USA.

- The patient-centered medical home (PCMH) is the example of chronic care for chronic disease to improve the outcome.\(^1\)

INTRODUCTION

- Many morbidities could be solved early, but they are not.
- More burden to health system (finance, manpower, time)
- USA: each admission lasted 5.1 days and cost per (patient or per admission) patient was $7,637.95.
- In five years, 37 patients spent 863 days in the hospital with a total cost of $1,283,176.83.\(^{(1)}\)
- Psychosocial on the patient, his family, and the community with rise unemployment, etc..

ORGANS COMPLICATIONS OF SICKLE CELL DISEASE

Retinopathy
Auditory impairment

Hepatomegaly
Gall stones
Cholestasis

Renal: enuresis, papillary necrosis, hyposthenuria.

Mesenteric occlusion

Priapism

Growth Retardation
Chronic Anemia

Stroke
Psychological implications

Heart failure
Pulmonary HT
Acute Chest S.

Splenomegaly

Delayed puberty
Reduced fertility
Obstetric complications

Skeletal:
Acute pain
Chronic pain
Osteonecrosis, Infection
BM expansion
Hand-Foot S.

Chronic Anemia
ARE WE BETTER?
CURRENT STATUS OF SCD IN SAUDI

- Distribution of SCD in the kingdom: certain regions
- No national or local registries
- No early diagnosis (neonatal screening)
- Limited access to medical care in many villages, and towns.
- Many patients undiagnosed.
- Guidelines developed and not popular or not known by the practitioners.
- Limited training for PHC doctors especially in highly prevalent areas.
- Medications and laboratory services limitations.
- 30-40% of adult medical bed occupancy by SCD patients.
- The system of care is not integrated one and drain resources.
Healthcare models used in cystic fibrosis and hemophilia (conditions that require lifelong multidisciplinary care) may provide helpful frameworks for SCD.

- It knowledgeable practitioners and a coordinated system of care throughout the patient’s lifespan.

- SCD patient live longer than the past, thereby need more care.

- Expand the services for unreached patients.
MODEL OF CARE

- There are several SCD model of care worldwide.

- Primitive services → center of excellence

- Scattered services → integrated system

- Simple → complicated
MINISTRY THOUGHTS

- Ministry of health studied health care system and in the VRO formed chronic diseases care unit.
- Model of care (MoC) unit studied the redesign the health delivery system.
- The aim is to have an integrated health care models.
- The regions were assigned different diseases
- SCD chosen in Eastern province (KFSH-D) is the base.
CHARACTERISTICS OF MOC

- To adopt
  - Integrated system
  - High yield benefits
  - Simple in application
  - Measurable process and outcome
  - Track patients
  - Reflect the patients need
  - Long term economic
- Sickle cell disease has a much higher prevalence in the Eastern Province than other areas.

- Eastern Province population have the sickle cell trait 24% and 1.2% have sickle cell disease.

- Therefore improving the sickle cell pathway is of strategic priority within the Eastern Province.
SOLUTIONS MODEL OF CARE
OVERVIEW: MODEL OF CARE FOR SAUDI

Physical wellbeing
Mental wellbeing
Social wellbeing

Chronic Care
Palliative Care
Preventive Care
Planned Care

Specialised Hospital Care
General Hospital Care
Primary Care
Virtual Care
Healthy Communities
Activated Person

Physical wellbeing
Mental wellbeing
Social wellbeing

Workforce
eHealth
Private-Sector Participation
Payment Mechanisms
Governance & Regulation
The system will support patients and their bereaved families during the last phase of their lives by providing them with compassionate care and empowering them to spend their last days where they are most comfortable.

The system will support people to manage their chronic conditions by providing integrated care and ease patient flow between care settings, and promoting care at the appropriate setting.

The system will support people when they have an urgent problem by providing needed treatment in the right place at the right time and supporting patients to return home and to community services.

The system will support women have a safe delivery and healthy infants by providing continuous support from pre-marriage and pre-conception to post-delivery and during early years development.

The system will support people to stay well – and get well again – by empowering them with knowledge and supporting them with healthy communities.

The system will support patients receive a great – and consistent - outcome for their planned procedures, by providing necessary, efficient, and quality care.
INTEGRATED SYSTEM

- Preventive initiatives
- Early detection
- Defined role for primary health care centers
- Hereditary blood disease centers
- Secondary care hospitals role
  - Emergency department
  - Admissions (inpatient services)
PREVENTION INITIATIVES

- **Three levels of prevention**
  1. **Public awareness**: the disease and the importance of follow up in the disease category
  2. **Pre-marital** screening initiatives and genetic counselling to inform prospective parents about the risks of their children inheriting the disease.
  3. **Preimplantation** genetic diagnosis (PGD) (i.e. screening for embryos that do not carry the sickle cell trait before implantation).
EARLY DIAGNOSIS INITIATIVES

- Screening levels
  - Neonatal screening esp. in high risk regions
  - Family members of high risk group (parents S trait, affected parent)
  - Population at risk and those not screened in the neonatal period
  - Primary healthcare centers (PHC) and inherited blood disease centers, and hospitals.
- Aims
  - Build national registries
  - Enroll the patients in comprehensive care program.
  - Social security support
PHCs roles:

- Diagnosis
- Health Education
- Genetic Counselling.
- Surveillance for chronic complications shared with IBDC through case coordinators.
- Refill medication (HU) and share follow up.
- Family doctors will be trained in SCD management complications and chronic pain.
- PHCs will be enhanced through the ‘Enhanced Primary Care’ intervention as part of the national Model of Care.
- Patient segmentation level 1 and 2
SCD patients can be classified based on severity of symptoms and developed complications and age group

Patients segmentation

Level 3: severe symptoms and major complications
- Require education, admissions in dedicated clinics and tertiary care procedures
- 1+ ACS (Achilles tendon calcificus)
- Cerebral infarcts
- Hepatic sequestration
- Splenic sequestration
- Frequent painful crises leading to hospitalization

Level 2: medium symptoms and few minor complications
- Mainly require education, regular check-ups in PHCs and ad-hoc home care assistance
- Medium painful crisis
- Bone necrosis
- Priapism
- Leg ulcer
- Congestive heart failure

Level 1: mild symptoms
- Mainly require education and regular check-ups in PHCs

1. This segment includes also patients with very mild/no symptoms at all.

Source:
- Evidence-based management of Sickle Cell Disease, NHLBI (2014)
- Development and validation of a pediatric severity index for sickle cell patients, American Journal of Hematology (2010)
INHERITED BLOOD DISORDER CENTERS

- **Roles**
  - Confirm sickle cell diagnosis
  - Keep up registry
  - Receive education for adult and pediatric families.
  - These centers could be co-located with secondary care hospitals to enable better integration.
  - Plan of care follow-ups will be carried out in PHCs with their family physician.
  - Patient assignment according to the segmentation (along with hospital SCD doctors)
  - Special procedures: TCD, US, X-rays, dexe scan, echocardiogram, ECG
  - Prescribe hydroxyurea and its plan and any other drugs
  - IBDC will keep annual visits for the patients for low risk, and special care consultations.
  - The clinic will allow easier case coordination of sickle cell patients with case coordinators working at the clinic full time, coordinating patients through all service layers and settings.
  - Patients should also be able to attend these clinics for pain episodes to alleviate the burden on EDs and receive pain management from staff trained to manage the disease, (a Later stage).
NATIONAL GUIDELINES

- The team will adopt or create comprehensive care program for SCD.

- ‘Virtual Self Care Tools’ may also be used to enable the patient and family to manage their condition at home, as well as education about potential complications and time table for appointments and consultations.
PATIENT ASSESSMENT, PATIENTS WITH COMPLICATIONS

- Chronic complications to IBDC or directly to hospital through case coordinators (AVN, gallstones, hypersplenism, iron overload)
- Acute complications to emergency department.
ADMISSIONS TO HOSPITALS

- The need for admissions are still to treat certain complications

- Acute complications:
  - Acute severe pain episode
  - ACS
  - Stroke
  - etc..

- Chronic complications:
  - Hips surgeries
  - Gallbladder stones
  - Splenectomy
TERTIARY CARE

- The role is limited to few referrals
- Stem cell transplantation.
- Other transplantations: liver, heart.
- Hip replacement: (secondary)
REQUIREMENTS

- Human Resources: case coordinators, family doctors, health educators, nurse practitioners, lab tech, IT tech.

- Ancillary Resources: networking, electronic files and single integrated health information system that everyone share the information, health education tools,

- Laboratory: Hb electrophoresis, kits, availability of tests and good utilization of resources. TCD, Ferritin, vit D

- Pharmacy: empower with medications HU, NSAID, Opioid.

- Development of SCD management guidelines.

- Training, training …
The pathway map for patients with sickle cell disease under the new Model of Care is shown below, note that numbers denote steps through the pathway and are detailed on the next few pages. Colours denote links to other systems of care, e.g. ‘Urgent Care’ for patients attending ED.

(0) Screening Initiatives
(1) Early Diagnosis
   - Virtual Education Tools
   - Chronic Disease Screening

Primary Care
   - Enhanced Primary Care

Secondary / Tertiary Care*
   (2) Emergency Department

Admission for acute pain episode
   - Pain Management if Necessary

Without complications (home discharge)

(2) Primary Healthcare Centre
   - Chronic Disease Screening
   - Route dependent on complication

(3) Inherited Blood Disorder Centre
   - Case Coordination
   - Direct walk-in to Inherited Blood Disorder Centre for more complex patients

Without complications

(4) Continuing Care at Home
   - Expert Patient
   - Enhanced Home Care
   - Continuing Care Services

(5) Outpatient Clinics
   - Referral to Specialist if required

(6) Inpatient Admission

*Referral to secondary or tertiary care depends on the severity of complications and will be based on clear national guidelines, tertiary referrals will usually be reserved for only the severe complex cases.
TESTING THE SYSTEM BY PILOT STUDY
SCD patients segmentation is based on symptoms and complications as well as age groups.

**Level 1: asymptomatic**
- Mainly require education and regular check-ups in PHCs

**Level 2: medium symptoms and few minor complications**
- Mainly require education, regular check-ups in PHCs and home care assistance

**Level 3: severe symptoms and major complications**
- Require education, admissions in dedicated clinics and tertiary care procedures

**Age group**
- Adults (15+ years)
- Pediatric (up to 14 years)

**More common in HbSS and HbSβ0 thalas. genotypes**

- 3 painful crisis per year
- 1+ ACS
- Pulmonary hypertension
- Chronic lung disease
- Cerebral infarcts
- Priapism
- Leg ulcer
- Hepatic/Intrahepatic sequestr.
- Splenic sequestr./Splenectomy
- Cholestasis
- Congestive heart failure
- Thrombotic crises
- Bone necrosis (late stage)
- Alloimmunization

- 1/2 painful crisis per year
- Bone necrosis (early stage)
- Stable proteinuria
- Asymptomatic gallstone
- Growth failure
- Hypertension
- Diabetes

**More common in HbSC and HbSβ+ -thalas. genotypes**

- Painful crisis per year
- ACS
- Pulmonary hypertension
- Chronic lung disease
- Cerebral infarcts
- Priapism
- Leg ulcer
- Hepatic/Intrahepatic sequestr.
- Splenic sequestr./Splenectomy
- Cholestasis
- Congestive heart failure
- Thrombotic crises
- Bone necrosis (late stage)
- Alloimmunization

**Source:** Evidence-based management of Sickle Cell Disease, NHLBI (2014); Development and validation of a pediatric severity index for sickle cell patients, American Journal of Hematology (2010); Evaluation of clinical severity in sickle cell disease, Journal of the National Medical Association (1983); A.T. Kearney

**Example Sample size**

<table>
<thead>
<tr>
<th>Level 1</th>
<th>Adults</th>
<th>Pediatric</th>
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<tbody>
<tr>
<td>Level 2</td>
<td>336</td>
<td>166</td>
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<tr>
<td>Level 3</td>
<td>224</td>
<td>119</td>
</tr>
<tr>
<td>Total</td>
<td>1,528</td>
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</tr>
</tbody>
</table>
WHY FUTURE PROJECT IS BETTER

1- it addresses issues from current status

2- it combine new interventions as well as cross-cutting interventions that is applicable to all systems of care.

3- it is patient centered as well as integrated
4- it is monitored by KPIs

It is sustainable financially.
Examples :
A- Emphasis on public awareness and health education for both families and patients who utilize virtual educational tools and virtual self care tools.
B- Focus on prevention: hydroxyurea available in PHCs to reduce pain and ED visit and addiction
C- PGD for high risk couples to produce well babies
D- Higher levels of efficacy and productivity
   Reduce ED visits so it will be utilized by different patients
   Through IBDC different providers are able to access patients files, prescriptions for continuity of care.
have a heart for sickle cell anemia