OPEN ACCESS

ISSN: 2997-321X

Sharma R, et al. (2024): 100056
Doi: 10.47991/2997-321X/AJCRR-112

Review Article

American Journal of Case Reports & Reviews

A Review of Comprehensive Care Model for Sickle Cell Disease Care

Rupam Sharma MD[1,2*], Leila Moosavi MD[1,2], Everardo Cobos MD[1,2,3]

[1]Department of Medicine, Kern Medical, Bakersfield, CA, USA
[2]David Geffen School of Medicine at UCLA, Los Angeles, CA, USA
[3]Division of Hematology/Oncology, Kern Medical, Bakersfield, CA, USA

[*]**Corresponding author:** Rupam Sharma MD, Kern Medical, Department of Internal Medicine; Bakersfield, CA USA. Tel: (661) 326-2000; Fax: (661) 862-7561; Email: sharma.rupam5@gmail.com

Citation: Sharma R, Moosavi L, Hodge S, Cobos E (2024) A Review of Comprehensive Care Model for Sickle Cell Disease Care. American J Cas Rep Rev: AJCRR-112.

Received Date: September 19, 2024; **Accepted Date:** September 30, 2024; **Published Date:** October 04, 2024

Abstract

Sickle cell disease (SCD), a genetic disorder leading to the mutation in hemoglobin beta chain. The prevalence of SCD is highest among people in Africa and high-resource countries such as the United States and Europe. Early management is mandatory, but newborn screening is not implemented everywhere. The development of care networks is essential to facilitate day to day care for these patients by defining the respective functions of nearby and highly specialized health care professionals working in close collaboration.

Keywords: *Sickle Cell Disease, comprehensive care model for sickle cell disease, sickle cell disease treatment, embedded care model.*

Introduction

Sickle cell disease (SCD) is a genetic disorder that causes a mutation in the hemoglobin (Hb) beta chain in which glutamic acid is substituted with valine at position 6 on chromosome 11. This mutation causes the polymerization of the Hb molecule that subsequently alters the shape of the red blood cell limiting its ability to deform as it passes through capillary beds resulting in small vessel occlusion in many organs. The prevalence of SCD is highest amongst people of Sub-Saharan Africa, South Asia, Middle East, and Mediterranean decent. [1] In the United States, most people with SCD identify as African American or of African ancestry with about 1 in 365 African American children being born with SCD with an estimated 100,000 people affected [1].

Although treatment for SCD has vastly improved over the last few decades including hydroxyurea, chronic blood transfusions and hematopoietic stem cell transplant, and recently introduced new agents (voxeletor, crizanlizumab, L-glutamine), many patients with SCD end up in the emergency department and frequent hospital admissions for pain crisis and other complications resulting in a lengthy and costly hospital admission. In a 2019 study conducted by the Healthcare Cost and Utilization Project, it was estimated that in 2016 there were over 134,000 SCD related inpatient hospital admissions with over 75% of these stays involving a pain crisis. [2]. They also found that the aggregate cost for inpatient admission for SCD totaled over $800 million with an average length of stay of five days, with one third of stays resulted in a 30-day readmission [2]. This highlights the importance of an evidence-based multidisciplinary comprehensive care approach to the treatment and management of patients with SCD to reduce morbidity, mortality, hospital admissions and health care costs to the patients and to the healthcare system.

Methods

Approval for this study was obtained from the Institutional Review Board of Kern Medical. Literature search was conducted on PubMed and Google Scholar. The following search terms were applied: Sickle cell disease, sickle cell diagnosis, sickle cell disease treatment, quality measures among sickle cell patients.

Discussion

The sickled cells are not able to deform as normal red blood cells do as they pass through capillary membranes causing the sickle cells to lead to ischemia and necrosis to any distal organ including liver, bone marrow, lungs, spleen, eyes, brain and skin. Commonly patients with SCD arrive to the emergency room complaining of severe pain which may be due to vaso-occlusive crisis, acute chest syndrome, priapism, osteonecrosis and aplastic crisis. Other potential complications of SCD include infections from encapsulated organisms secondary to asplenia, pulmonary hypertension, stroke, renal failure, as well as proliferative retinopathy [1].

The literature on initiatives surrounding comprehensive care among SCD patients have examined the implementation of standardizing care pathways or use of individualized care plans in the emergency department and have shown to improve pain management, however most of these projects have not decreased admission rates [5-6]. This review aims to describe the different

care models used presently in taking care of sickle disease patients including our developed Kern Medical model.

The Kern Medical model of care for SCD patients is a blend of the current three comprehensive care models: Classic comprehensive model, Embedded care model and the specialized home care model.

Classic comprehensive Model

The classic comprehensive model is a coordinated, team based SCD care model with dedicated clinical workers and space. This model provides access to sickle cell specialist, social worker, community health worker, advanced nurse practitioners and others focused in the particular local population. This model also provides access to an infusion center, blood bank, radiology, laboratory services and other disciplines required in care of SCD patients [1].

Embedded Care Model

The Embedded Care Model is instituted in models without dedicated space and staff. It 'embeds' itself within a larger, more financially feasible institution. In this model, the clinical space is shared thereby reducing costs and improving efficiency. This model encompasses team-based care and shared personnel though led by a sickle cell specialist. Additionally, this model also provides access to laboratory services, radiology, blood bank and other disciplines required in care of SCD patients [1].

Specialized Home Care Model

The specialized home care model is unique in a way that it provides medical home including primary care services within the comprehensive sickle cell care model. This model was designed to specifically target the complications of this disease while providing comprehensive care with the team lead being the primary care physician (PCP). The PCP in conjunction with the sickle cell specialist provides collaborative and simultaneous care to the patients. This model also provides access to laboratory services, radiology, blood bank and other services required in care of SCD patients [1].

Kern Medical Model

The model of care implemented here at Kern Medical is a blend of the above mentioned three models of cares. Our clinic is in a setting of a shared space within the oncology infusion center, making it more financially feasible. Our team comprises of a lead sickle cell specialist, pharmacist, pain specialist, primary care physician, behavioral health staff, case manager, nurses and a community health worker.

Few years prior to the implementation of our multidisciplinary sickle cell center sickle cell patients were frequently coming to Bakersfield with no medical care. Based on the initial needs assessment, majority of our patients in Kern County had not been followed by a hematologist previously. Additionally, genotype testing, starting on disease modifying agents, or even regular PCP follow up were not standard of care.

Regular hematology follows up have shown to decrease acute care utilization, health care costs, and improved quality of life [3-4]. An important parameter to follow is the hemoglobin F level along with a formalized recommended individualized pain regimen. A sickle cell provider can optimize disease modifying agents to prevent sickle cell organ damage. Frequent outpatient follows up can decrease worsening disease, which will decrease acute care utilization. With a comprehensive sickle cell center, individuals can be not only managed medically but also have their social barriers to health addressed as well. If we start to manage individuals who are high acute care utilizers, we would improve the quality of care of the patients and decrease our financial burden. The goal of the comprehensive sickle cell clinic is to provide a medical home to all sickle cell patients in Kern County.

Conclusion

In conclusion, though treatment for SCD has vastly improved over the last few years, many patients with SCD rely on the emergency department as their mode of primary care. From our review it was obvious that the quality indicators were poor prior to the implementation of our clinic and highlighted the need for a SCD clinic. The importance of a comprehensive care approach to the treatment and management of patients with SCD is essential.

Ethics Approval

Ethical approval to was obtained from the Kern Medical Institutional Review Board.

Acknowledgements: This case was presented orally at the American Federation of Medical Research's Western Conference, Carmel, CA in January 2022 and the Southern San Joaquin Valley Research Forum in May 2022.

Funding: The author(s) received no financial support for the research, authorship, and/or publication of this article.

Declaration of Conflicting Interests: The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Authors contribution: RS wrote the initial draft of the manuscript. LM and EC revised and reviewed the manuscript.

References

1. Sedrak A, Kondamudi NP (2020). Sickle Cell Disease. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing; August 16, 2020.
2. Fingar KR (IBM Watson Health), Owens PL (AHRQ), Reid LD (AHRQ) et al. (2019) Characteristics of Inpatient Hospital Stays Involving Sickle Cell Disease, 2000-2016. HCUP Statistical Brief #251. September 2019. Agency for Healthcare Research and Quality, Rockville, MD. www.hcup-us.ahrq.gov/reports/statbriefs/sb251-Sickle-Cell-Disease-Stays-2016.pdf.
3. Balsamo, L., et. al. (2019). Improving Care for Sickle Cell Pain Crisis Using a Multidisciplinary Approach. *Pediatrics*, *143*(5). doi:10.1542/peds.2018-2218.
4. Powell, R. E., et al. (2017). A Multidisciplinary Approach to Impact Acute Care Utilization in Sickle Cell Disease. *American Journal of Medical Quality*, *33*(2), 127-131. doi:10.1177/1062860617707262.

5. Kato-Lin YC, Krishnamurti L, Padman R, Seltman HJ (2014) Does e-pain plan improve management of sickle cell disease associated vaso-occlusive pain crisis? Amixed methods evaluation. Int J Med Inform. 2014;83(11):814–824.

6. Krishnamurti L, Smith-Packard B, Gupta A, et. al. (2014) Impact of individualized pain plan on the emergency management of children with sickle cell disease. Pediatr Blood Cancer. 2014;61(10):1747–1753.