SICKLE CELL ELECTIVE ROTATION

Principal Preceptor: **Dr. Adetola Kassim**

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Rotation Site(s):

Browning in-patient service – 3.5 days/week

Benign Heme consultation service – 3.5 days/week

Outpatient sickle cell clinic – Wednesday mornings, all day Friday

Overall Learning Objectives:

At the conclusion of this elective, residents should be able to:

Knowledge:

- Understand the genetics and pathophysiology of sickle cell disease, as well as psychosocial factors, poverty and other socioeconomic factors, how PCPs fit into the care model
- Demonstrate understanding of the pathophysiology and treat of acute pain episodes and chronic pain (eg, avascular necrosis, musculoskeletal issues)
- Recognize and manage acute complications in SCD, including: acute chest syndrome, priapism, splenic sequestration, abdominal pain, vision changes
- Recognize and manage chronic complications in SCD, including: Heart, lung, and kidney issues, leg ulcers, CNS complications – strokes, silent strokes, and cognition.
- Recognize other problems that may exacerbate SCD such as diabetes, tobacco use, hypertension
- Identify disease modifying therapies: Medications (hydroxyurea, L-glugtamine (Endari), voxeletor (Oxbryta), crizanlizumab (Adakveo)), chronic transfusion therapy
- Identify curative therapies for SCD: Bone marrow transplant and Gene therapy
- Feel comfortable with PCP management of SCD
- Resources for the PCP: Guidelines (2014 NHLBI, 2020 ASH), checklists, patient resources, the patient portal, apps
- Demonstrate knowledge of supportive care: Vaccines and screening, depression, women's health, hypertension, diabetes, vitamin D and bone health, sleep, acute issues

Skills:

- Demonstrate indications and exclusions, complications, efficacies, dosing, and side effects/toxicities of how to manage hydroxyurea
- Recognize patients' perspective of disease, including both positives and negatives of the care they receive
- Exhibit post-transplant knowledge for the PCP, including how to talk to patients about curative therapies, long term complications and side-effects
- Understand about home pain action plan and ED management approaches
- Know how to utilize available resources for psychosocial support of patients with SCD
- Utilize the medical home model, and gain knowledge of the role of the nurse case manager in coordination of care
- Recognize the impact of implicit and explicit bias on care of patients with SCD

The rotation will include:

-7 half-days per week on the inpatient Browning or Benign Hematology consult service

-3 half-days in the outpatient sickle cell clinic

-Afternoon didactic lectures

-Conferences: Sickle cell pathways conference 8.00 am – 9.00 am weekly

Benign Heme conference every Friday (12 pm – 1.00pm)

Time Off:

Per department policy. Residents will have evenings and weekends off.

Conferences:

Sickle cell pathways conference 8.00 am – 9.00 am weekly

Benin Heme conference every Friday (12 pm – 1.00pm)

SAMPLE Weekly Summary grid for Rotation:

	Monday	**Tuesday**	**Wednesday**	**Thursday**	**Friday**
8-9 am	Inpt	Inpt	Inpt	Inpt	Pathway conference
9-12 pm	Inpt	Inpt	Clinic	Inpt	Clinic
12 – 1 pm	Inpt	Inpt	Inpt	Inpt	Benign Heme conference
1-4.30pm	Inpt	Inpt	Inpt	Inpt	Clinic

Suggested Readings:

1. Barbara P. Yawn, George R. Buchanan, Araba N. Afenyi-Annan, et. al. Management of Sickle Cell Disease Summary of the 2014 Evidence-Based Report by Expert Panel Members. JAMA. 2014;312(10):1033-1048.

2. Stella T. Chou, Mouaz Alsawas, Ross M. Fasano. American Society of Hematology 2020 guidelines for sickle cell disease: transfusion support. Blood Adv. 2020 Jan 28;4(2):327-355.

3. Robert I. Liem, Sophie Lanzkron, Thomas D. Coates et al. American Society of Hematology 2019 guidelines for sickle cell disease: cardiopulmonary and kidney disease. Blood Adv. 2019 Dec 10;3(23):3867-3897.

4. M. R. DeBaun, L. C. Jordan, A. A. King et. al. American Society of Hematology 2020 guidelines for sickle cell disease: prevention, diagnosis, and treatment of cerebrovascular disease in children and adults. Blood Adv. 2020 Apr 28;4(8):1554-1588.

5. Amanda M. Brandow, C. Patrick Carroll, Susan Creary et al. American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain. Blood Adv. 2020 Jun 23;4(12):2656-2701.

6. Ataga KI, Kutlar A, Kanter J, et al. Crizanlizumab for the prevention of pain crises in sickle cell disease. *N Engl J Med*. 2017;376(5):429-439.

7. Quinn CT. l-Glutamine for sickle cell anemia: more questions than answers. Blood 2018; 132:689.

8. Niihara Y, Miller ST, Kanter J, et al. A Phase 3 Trial of l-Glutamine in Sickle Cell Disease. N Engl J Med 2018; 379:226.

9. Niihara Y, Zerez CR, Akiyama DS, Tanaka KR. Oral L-glutamine therapy for sickle cell anemia: I. Subjective clinical improvement and favorable change in red cell NAD redox potential. Am J Hematol 1998; 58:117.

10. Niihara Y, Macan H, Eckman JR, et al. L-Glutamine therapy reduces hospitalization for sickle cell anemia and sickle β°-thalassemia patients at six months - A phase II randomized trial. Clin Pharmacol Biopharm 2014; 3:116.

11. Minniti CP. l-Glutamine and the Dawn of Combination Therapy for Sickle Cell Disease. N Engl J Med 2018; 379:292.

12. Vichinsky E, Hoppe CC, Ataga KI, et al. A Phase 3 Randomized Trial of Voxelotor in Sickle Cell Disease. N Engl J Med 2019; 381:509.

13. Blyden G, Bridges KR, Bronte L. Case series of patients with severe sickle cell disease treated with voxelotor (GBT440) by compassionate access. Am J Hematol 2018.

14. Oksenberg D, Dufu K, Patel MP, et al. GBT440 increases haemoglobin oxygen affinity, reduces sickling and prolongs RBC half-life in a murine model of sickle cell disease. Br J Haematol 2016; 175:141.

15.Estepp JH. Voxelotor (GBT440), a first-in-class hemoglobin oxygen-affinity modulator, has promising and reassuring preclinical and clinical data. Am J Hematol 2018; 93:326.

16.Kutlar A, Kanter J, Liles DK, et al. Effect of crizanlizumab on pain crises in subgroups of patients with sickle cell disease: A SUSTAIN study analysis. Am J Hematol 2019; 94:55.

17.https://www.accessdata.fda.gov/drugsatfda_docs/label/2019/761128s000lbl.pdf (Accessed on November 19, 2019).

18. DeBaun MR. Initiating adjunct low-dose hydroxyurea therapy for stroke prevention in children with SCA during the COVID-19 pandemic. Blood. 2020;135(22):1997-1999.*

19. Robert Sheppard Nickel, Stefanie Margulies, Brittany Frazer, Naomi L. C. Luban, and Jennifer Webb. Combination dose-escalated hydroxyurea and transfusion: an approach to conserve blood during the COVID-19 pandemic*