OVERVIEW OF HEALTHCARE ADMINISTRATIVE, RADIOLOGY AND MEDICAL LABORATORY ROLES IN MANAGEMENT OF SICKLECELL DISEASE PATIENTS

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Abstract:
While advancements in medical care have made it possible for juvenile sickle cell disease patients to reach adulthood, the management of the disease in adult sickle cell disease patients, continues to be difficult due to the higher rates of mortality and comorbidities. In addition, adults are more likely to be hospitalized than pediatric patients because they are more likely to have received chelation or transfusions and to have signs and symptoms of classic chronic diseases. Establishing the right diagnosis of sickle cell anemia (SCA) at an early age, preferably during the newborn period, is the first step in beginning the appropriate care of this condition. The roles of health care adminstration, together with radiology and medical laboratory teams are very crucial in management of those patient. The early commencement of prophylactic vaccines against pneumococcal and penicillin, which help prevent overwhelming sepsis, is made possible by the identification of afflicted infants through neonatal screening programs. Ongoing education of families helps to encourage the early recognition of difficulties that are caused by the disease, which in turn enables timely and appropriate medical diagnosis and therapeutic response. In order to provide comprehensive care, periodic evaluations by trained professionals are helpful. These evaluations include transcranial Doppler tests, which are used to detect children who are at risk for primary stroke.

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Introdution:

The erythrocytes of the body, which are responsible for the transportation of oxygen throughout the body and contain hemoglobin, can become crescent or sickle-shaped as a result of a mutation in the HBB gene, which is the cause of sickle cell disease (SCD), a dangerous disorder that is hereditary. Consequently, an aberrant beta-globin molecule known as hemoglobin S (HbS) is formed as a consequence of this issue. HbS is a result of an abnormality in nucleotide substitution, which leads to the substitution of valine for glutamate in hemoglobin. It is responsible for the replacement of both beta-globin subunits because it is the cause of the anomaly. Red blood cells undergo a conformational change as a result of the production of hemoglobin S, which causes them to become sickle-shaped rather than spherical, resulting in increased tensile strength and stickiness [2]. Because of these modifications, they have a tendency to aggregate and cling to the walls of small blood arteries, which obstructs blood flow and causes a variety of health problems in both children and adults, including sickle cell anemia [3].

Additional issues that are connected with sickle cell disease are more common in children than in adults. Some of these conditions include dactylitis and hand-foot syndrome, growth retardation, delayed sexual maturation, being underweight, increased susceptibility to infections, aplastic crisis, vaso-occlusive events, splenic sequestration and infarction, strokes, and injuries to the brain such as a silent cerebral infarct. Acute chest syndrome is another condition that can occur. Damage to the lungs, heart, and kidneys, in addition to damage to the eyes, are some of the other issues that can occur in adolescents and adults. However, this list is extensive. There is also the possibility of developing priapism, in addition to gallstones, leg ulcers, deep vein thrombosis, pulmonary embolism, neurological impairments, chronic hemolysis, pulmonary artery hypertension, and issues that arise from receiving frequent blood transfusions [4].

Because of the varied degrees of complexity that these problems can have, some of which can become life-threatening, there are a variety of therapeutic alternatives that can assist patients in alleviating the extreme agony that they are experiencing. Moreover, treatment modalities not only contribute to the improvement of a patient's well-being, but they also offer potential means of assisting in the prevention of the recurrence of complications and the reduction of death rates [5].

Thankfully, significant progress has been made over the course of the past four decades, and improved management practices have resulted in a change from the previously pessimistic view. Despite the complexity and multifactorial pathophysiology of vaso-occlusion, relatively straightforward measures have greatly improved outcomes for children with SCA: early identification by neonatal screening programs; education of parents and patients about medical complications and early recognition; preventive measures with prophylactic penicillin and pneumococcal immunizations; aggressive treatment of acute VOEs including hydration, analgesics, antibiotics, and transfusions; screening programs for early signs of organ damage, especially primary stroke risk using transcranial Doppler (TCD) examinations; and therapeutic intervention with transfusions, hydroxyurea, or stem cell transplantation. There is evidence that children who receive medical care in comprehensive care programs have survival rates that range from 95% to 99% of their lives into adulthood. Even while screening programs and anticipatory guidance are less standardized for adults with SCA, they are nevertheless extremely important. Furthermore, the benefits of preventative therapy that utilizes hydroxyurea are even more convincing at this point [5,6].

Review:

The presence of homozygous hemoglobin S (Hb S) is the defining characteristic of sickle cell anemia (SCA), which is characterised as an autosomal recessive disease that is inherited. A single nucleotide mutation is responsible for this condition, which occurs when glutamic acid is substituted for valine in the sixth position of the β-globin gene rather than valine. During hypoxic conditions, the red blood cell undergoes a change in structure that causes it to become sickled. This change in structure is responsible for restricting circulation, which in turn causes the capillaries to get blocked, which ultimately leads to the premature death of the cell [7].

There is a wide range of clinical manifestations linked with SCA, ranging from mild variants, which are essentially no symptoms at all, to severe versions that are associated with significant death rates.3) Clinical signs typically reveal themselves at the age of three months, which coincides with a decline in the concentration of fetal hemoglobin (Hb F).2. The majority of systems are susceptible to vaso-occlusive processes, which could potentially lead to failure of multiple systems [8].

At this time, there is no cure that can be considered definitive for those who have SCA. Treatments that are currently available are mainly concerned with the management of symptoms; they do not modify the normal progression of the disease. Hydration, the avoidance of infections, the management of pain, the provision of appropriate nourishment, and the taking of precautions against adverse weather conditions are the components that make up these therapies. As a result, additional medicines are required in order to minimize problems without exposing patients to the higher morbidity and mortality that is associated with very aggressive procedures such as hematopoietic stem cell transplantation (HSCT) [8].

There is currently just one medical approach that has been shown to be effective in treating patients who experience frequent symptoms connected to SCA, and that is hydroxyurea (HU). The administration of HU has been demonstrated to raise the levels of Hb F, enhance the concentrations of hemoglobin and the mean corpuscular volume, and decrease the number of reticulocytes. Not only does the treatment lessen the expression of adhesion molecules, but it also lessens the quantity of receptor proteins that are found on endothelial cells. This is yet another beneficial reaction that the medication has. Because of this, HU reduces the amount of vascular adhesion, which in turn contributes to a reduction in the number of vaso-occlusive panic attacks [9].

It is possible that the early diagnosis of affected people, prior to the onset of signs and symptoms of disease, is the most important part of optimizing the care of SCA. Within the first few years of life, a significant number of newborns pass away unexpectedly due to bacterial sepsis or acute splenic sequestration crisis (ASSC) if they do not receive an early diagnosis and adequate treatment. This is because SCA frequently works as a rapid and undetected killer. Even before families or medical providers are aware that their infants have sudden cardiac arrest (SCA), deadly consequences can sometimes materialize [9].

The landmark multicenter double-blind placebo-controlled PROPS trial demonstrated that penicillin prophylaxis considerably reduced the risk of bacteremia and death [10]. This was accomplished in light of the fact that infants with SCA have a dramatically elevated risk of bacterial sepsis. Justification for newborn screening for sudden cardiac arrest (SCA) was established by this straightforward intervention, which enabled afflicted newborns to be identified shortly after birth and enabled life-saving prophylactic antibiotic medication to be administered. There has been a significant decrease in morbidity and mortality, particularly in the first five years of life, which demonstrates that early identification of SCA through neonatal screening programs has contributed to the improvement in survival rates [10]. Newborn screening programs in the United States, Jamaica, and Europe have documented the usefulness of early identification of SCA.

Both children and adults who have sickle cell disease require primary care as well as specialty care services continually throughout their whole lives. It is common for the unique components of care to result in an unanticipated requirement for urgent and emergency care to handle pain, fever, and neurologic symptoms. On the other hand, the needs for care linked to screening, early detection, and the management of chronic sequelae are more predictable. The provision of a wide range of specialty services is necessary. These services include multidisciplinary expert providers, medical subspecialists, primary care physicians (PCPs), surgeons, anesthesiologists, radiologists, social workers, mental health specialists, care coordinators, and community health workers (CHWs). As a result of their support for patient and community education, provision of counseling, and elimination of barriers to care through the provision of services ranging from transportation to care coordination, community-based organizations (CBOs) are an indispensable component of this care. It is essential to provide care that is of a high quality and that is in accordance with the six different aspects of health care quality. Furthermore, it is essential for those who give medical care to comprehend and take into account the unfavorable societal views that are held about members of the SCD population. These views include racism, stigma, and a variety of stereotypes, such as the presumption that someone who is living with chronic pain is looking for drugs. Not only do these perspectives affect the manner in which care is offered, but they also affect who delivers that care and where it is provided. Despite the fact that these nuances are frequently overlooked in published literature, the committee is of the opinion that they should be taken into consideration when determining the most effective means of providing high-quality medical treatment to those who have sickle cell disease [11].

Bright Futures publications outline the information that should be covered during well-child visits. This content includes health supervision,

anticipatory guidance, monitoring of both physical and psychosocial development, as well as appropriate periodic screening and vaccines. In addition, the Early Periodic Screening, Diagnosis, and Treatment (EPSDT) benefit stipulates that children under the age of 21 who are enrolled in Medicaid must be provided with screening and treatment services. Although the two methods do have some similarities, the EPSDT places a greater emphasis on dental screening and treatment, as well as hearing and vision screening [12]. A different approach than that of ordinary pediatric health care is required in order to provide the extensive range of services that are required by children who have sickle cell disease (SCD). One of the models of care that is available for children with sickle cell disease is the medical home, which is a model that was developed by the American Academy of Pediatrics (AAP). This model is available for children who are unable to access specialized sickle cell care centers. In an ideal world, a medical home would provide care that is not only accessible but also continuous, comprehensive, family-centered, coordinated, compassionate, and culturally effective to each and every child and adolescent. The term "pediatric medical home" refers to a partnership that is centered on the family and operates within a community-based system. This partnership offers continuous treatment and appropriate funding in order to support and maintain optimal health outcomes. Beginning at birth and continuing through the transition into adulthood, medical homes provide healthcare that is preventative, acute, and chronic. An integrated health system is made possible by a medical home, which promotes the formation of an interdisciplinary team consisting of patients and their families, primary care physicians, specialists and subspecialists, hospitals and other health care institutions, public health, and the community [12].

Conclusion:

Treatment and follow-up for sickle cell anemia must be provided by a team consisting of multiple professionals. Hydroxyurea is currently being considered as a potential treatment approach. Patients who use this medication see fewer problems and an improvement in their laboratory results. Employees in the healthcare industry, such as those working in administration, radiography, and medical laboratories, are increasingly being acknowledged as valuable contributors to the enhancement of health care and health outcomes for a spectrum of chronic disorders. Lay counseling, social support, and assistance in navigating health systems and resources are all services that community health workers are able to give. Community health workers should make an effort to match their social and cultural practices with those of the population they serve. This is an essential component of community health worker intervention. Despite the fact that it has been demonstrated that interventions carried out by community health workers can improve patient-centered outcomes in marginalized settings, these treatments have not been examined with sickle cell disease. There is evidence from other disease areas that shows that the intervention of community health workers would also be useful for these patients of the same condition. The disease known as sickle cell is a complicated condition that presents a variety of obstacles to the provision of multimodal treatment at the person, family/friend, clinical organization, and community levels. Disparities in health care, including access, delivery, services, and cultural incompatibilities between clinicians and families, make the delivery of care more difficult. The approaches that are now being utilized either do not sufficiently address or provide insufficient control of symptoms, particularly pain, which leads to a decline in quality of life and an increase in the cost of medical care.

References:

1. Shankar SM, Arbogast PG, Mitchel E, Cooper WO, Wang WC, Griffin MR. Medical care utilization and mortality in sickle cell disease: a population-based study. *Am J Hematol*. 2005;80(4):262–270.
2. Lanzkron S, Haywood C, Jr., Segal JB, Dover GJ. Hospitalization rates and costs of care of patients with sickle-cell anemia in the state of Maryland in the era of hydroxyurea. *Am J Hematol*. 2006;81(12):927–932.
3. Kauf TL, Coates TD, Huazhi L, Mody-Patel N, Hartzema AG. The cost of health care for children and adults with sickle cell disease. *Am J Hematol*. 2009;84(6):323–327.
4. Mvundura M, Amendah D, Kavanagh PL, Sprinz PG, Grosse SD. Health care utilization and expenditures for privately and publicly insured children with sickle cell disease in the United States. *Pediatr Blood Cancer*. 2009;53(4):642–646.
5. Raphael JL, Dietrich CL, Whitmire D, Mahoney DH, Mueller BU, Giardino AP. Healthcare utilization and expenditures for low income children with sickle cell disease. *Pediatr Blood Cancer*. 2009;52(2):263–267.
6. Brousseau DC, Owens PL, Mosso AL, Panepinto JA, Steiner CA. Acute care utilization

and rehospitalizations for sickle cell disease. *JAMA.* 2010;303(13):1288–1294.
7. Lanzkron S, Carroll CP, Haywood C., Jr. The burden of emergency department use for sickle-cell disease: an analysis of the national emergency department sample database. *Am J Hematol.* 2010;85(10):797–799.
8. Yusuf HR, Lloyd-Puryear MA, Grant AM, Parker CS, Creary MS, Atrash HK. Sickle cell disease: the need for a public health agenda. *Am J Prev Med.* 2011;41(6 Suppl 4):S376–383.
9. Raphael JL, Mei M, Mueller BU, Giordano T. High resource hospitalizations among children with vaso-occlusive crises in sickle cell disease. *Pediatr Blood Cancer.* 2012;58(4):584–590.
10. Anderson N, Bellot J, Senu-Oke O, Ballas SK. Characteristics of acute care utilization of a Delaware adult sickle cell disease patient population. *Popul Health Manag.* 2014;17(1):60–65.
11. Redha NA, Mahdi N, Al-Habboubi HH, Almawi WY. Impact of VEGFA -583C > T polymorphism on serum VEGF levels and the susceptibility to acute chest syndrome in pediatric patients with sickle cell disease. *Pediatr Blood Cancer.* 2014;61:2310–2.
12. Kayle M, Docherty SL, Sloane R, Tanabe P, Maslow G, Pan W, et al. Transition to adult care in sickle cell disease: A longitudinal study of clinical characteristics and disease severity. *Pediatr Blood Cancer.* 2019;66:e27463.