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Sickle - Cell Disease

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ABSTRACT

Sickle cell disease is a common inherited, multisystem, monogenic disorder of red blood cells (erythrocytes) caused due to polymorphic changes in hemoglobin. The most commonly known condition; Sickle cell anemia wherein there aren't enough healthy RBCs to carry adequate oxygen throughout the body is considered to be a common form if Sickle-cell disease. Other type of Sickle cell disease is Hemoglobin SC disease (HbSC) caused due to inheritance of beta s and beta c alleles. The third type of such disease is HbS thalassemia caused due to beta-thalassemia mutation in the beta-globin gene leading to Sickle hemoglobin (HbS). Polymerization of HbS due to presence of fetal hemoglobin in the erythrocytes that in turn reduces the concentration of HbS which becomes the prominent determinant to check the severity of the disease. Reduced concentrations of HbS also reduce hemolysis that prevents acute vaso- occlusion. This pain is caused as the irregular shaped RBCs and WBCs get entrapped in the small blood vessels causing vascular obstruction and tissue ischemia. HbS polymerization can also lead to hemolytic anemia which is a state where in rate of RBC destruction is faster than formation; such patients are likely to develop vasculopathy. During the process of hemolysis, hemoglobin is released into plasma that inhibits endothelial nitric oxide signaling causing endothelial cell dysfunction. Hemolysis is also associated with formation of erythrocyte microvesicles that acts as a activator of tissue factor. Malaria is considered to cause HbS. Sickle cell disease is found to be highest in the African continent mostly affecting the new borns. The cause of deaths is hug in Africa due to poor diagnostic facilities. Measures taken against H influenzae and S pneumoniae that is profoundly detected in African children with Sickle cell disease can help reduce the disease proximity. Implementation of early life screening can thereby be effective in this case.

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The disease can be diagnosed by analysis of hemoglobin content of individual height techniques like protein electrophoresis and chromatography. Antenatal screening for women and neonatal screening help facilitate early diagnosis and treatment. Oral, less toxic drug, Hydroxycarbamide is chosen as a treatment against Sickle cell disease as it facilitates in increasing hemoglobin concentrations and also administers other problems associated with Sickle cell anemia in both adults and children. The drug even has some side-effects including male subfertility.

Complications may arise in and cause of Sickle cell disease. Blood transfusions are associated with heavy iron content and iron retainance in liver of patient which causes a need for iron removal by oral iron chelator like defrasirox. Similarly other complications that arise with Sickle cell anaemia is acute pain which can be administered by certain drugs but can be fatal. Hence no specific treatment has yet been found. Other rising risk include bacterial infections along with impaired splenic functions, micronutrient deficiency especially in children. This can be treated by penicillin and making use of conjugate vaccines. Strokes associated with neurological origin are often seen in children due to Sickle cell disease. It requires a regular blood transfusion to prevent stroke. Tran's cranial Doppler screening can be applied for early detection and timely prevention of stroke. The disease can silently affect the frontal lobes of the brain in adults and children leading to cognitive impairment and intracranial bleeding. Neurosurgical treatment is suggested but positive outcome is poor. Another major complication in such patients leading to hospitalization is acute chest syndrome which is a severe lung injury. The treatment involves broad spectrum antibiotics, bronchodilators and oxygen. In case of reduction of hemoglobin, blood transfusion is performed which can be avoided using drugs like Dexamethasone? One of the increasing complications of Sickle cell disease in teenagers and adults is pulmonary hypertension. Echocardiography can be used to measure the pressure in pulmonary artery greater than 35mm considered safe. Occurrence of pulmonary hypertension is comparatively higher than other associated diseases but it can be treated by administration of drug Hydroxycarbamide with regular blood transfusions. Idiopathic pulmonary hypertension is a severe type that can be treated using prostaglandin based therapy. Heart disease is also a complication seen dye to diastolic and systolic dysfunction . Patient suffering from pulmonary disease as well as diastolic dysfunction are at a higher risk of deaths. Similar to heart complications patients also come across renal complications accompanied with Sickle cell disease as the HbS tends to polymerize in the renal medulla. This also causes glomerular hyper filtration and tubular dysfunction this ultimately is associated with anemia. Some other renal manifestations hematuria, nocturnal enuresis. The treatment focused is significant use of Hydroxycarbamide. Thus it is studied that Sickle cell disease is seen to be causing a multisystem problems.

Development in the areas of stem cell transplantation and gene therapy are more likely to be promising in the near future along with pharmaceutical approaches which would benefit a large number of patients across globe.