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Heamostatic and genetic predisposing factors for stroke in children with sickle cell anemia

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Sickle cell disease (SCD) is a group of genetic disorders characterized by the production of the abnormal hemoglobin S (HbS). Sickle cell anemia (SCA) is the most common type of SCD and represents the homozygous form, in which the individual inherits a double dose of the abnormal gene that codes for hemoglobin S. This study was done to detect cases of silent and clinically overt strokes in children with sickle cell anemia (SCA) and examine predisposing factors for stroke development. The study included 20 children with clinically and hematologically confirmed SCA and 10 controls. They were divided into two groups, group I; included 10 steady state cases and group II; included 10 cases with thrombotic crisis. All subjects were subjected to full clinical examination, measurements of plasma level of: fibrinopeptid A (FPA), thrombin-antithrombin III (TAT), fibrin degradation product (D-dimer) and serum level of platelet endothelial cell adhesion molecule-1 (PECAM-1), and analysis of the ACE gene polymorphism by polymerase chain reaction (PCR). Patients were further subjected to Brain computed axial tomography (CT) scan and/or magnetic resonance imaging (MRI) as well as electro-encephalographic studies (EEG). Silent ischemic brain infarction as evidenced by CT scan and/or MRI was present in one patient in group I (10%) and one patient in group II (10%). On the other hand, two patients in group II (20%) showed clinically overt strokes. Thus, 4 children had silent or clinically overt stroke and the remaining 16 were non-stroke cases. Laboratory results showed that the levels of FPA, TAT, D-dimer and PECAM-1 were significantly elevated in SCA patients both in the steady and crisis states as compared to control, with more evident significant elevation in group II (thrombotic crisis) as compared to group I (steady state). The stroke group showed significant elevation; FPA, TAT, D-dimer and PECAM-1 as compared with non-stroke group. The PCR results showed that the frequencies of both DD genotype and D allele of ACE gene in the thrombotic crisis are significantly higher than in the control group and that all stroke children are of DD genotype. In conclusion, significant increase in FPA, TAT, D-dimer and PECAM-1 levels as well as the presence of ACE D allele of the ACE gene are significant predisposing factors for stroke in children with SCA. Regular follow-up by thorough neurological examination and neuro-imaging studies for early detection of silent brain infarction is recommended.