**Appendix 1:** DFConhecimento instrument

|  |
| --- |
| **SICKLE CELL DISEASE KNOWLEDGE SURVEY** |
| 1. Among the diseases screened by the Neonatal Screening Program, which one has the highest incidence in the Brazilian population?( ) Congenital hypothyroidism ( ) Phenylketonuria**(answer) Sickle cell disease**( ) Cystic fibrosis( ) Biotinidase deficiency( ) Congenital adrenal hyperplasia( ) Don’t know2. When discussing Sickle Cell Disease, it is CORRECT to state that( ) It occurs among the black population in Brazil( ) It is one of the rarest genetic disorders in Brazil and worldwide.**(answer) It is characterized by the predominance of hemoglobin S in red blood cells**( ) Its SS genotype is a mild form of the disease( ) Don’t know3. Which one is the Sickle Cell Anemia Genotype?( ) Hb SC( ) Hb AS**(answer) Hb SS**( ) Hb SD( ) Hb AA( ) Don’t know4. When discussing persons with sickle cell trait, it is CORRECT to state that they( ) Need monitoring and hematological treatment( ) Suffer from anemia**(answer) Do not have a disease. Have a relatively common and clinically benign condition**( ) Are not advised to practice physical activity ( ) Don’t know5. All of the following are features of sickle cell disease EXCEPT FOR( ) Jaundice( ) Frequent infections( ) Hemolytic anemia**(answer) Iron deficiency anemia**( ) Biliary calculus( ) Don’t know6. All of the following manifestations are caused by vaso-occlusion EXCEPT FOR( ) Pain cramps( ) Priapism( ) Leg ulcers( ) Retinopathy( ) Osteonecrosis**(answer) Hyperglycemia**( ) Hand-foot syndrome (Dactylitis)( ) Stroke ( ) Don’t know 7. All of the following conditions favor the sickling of erythrocytes EXCEPT FOR( ) Dehydration( ) Strenuous physical activity( ) Exposure to cold**(answer) Being overweight**( ) Emotional stress( ) Don’t know8. All of the following signs are considered warning signs in sickle cell disease EXCEPT FOR( ) Fever( ) Enlarged spleen and liver( ) Coughing or shortness of breath( ) Abdominal pain**(answer) Feet and leg Itching**( ) Worsening in pallor of skin and mucous membranes( ) Nausea / vomiting( ) Neurological deficit( ) Don’t know9. All of the following drugs are used to prevent and/or treat sickle cell disease and its intercurrences EXCEPT FOR( ) Penicillin( ) Folic acid**(answer) Ferrous sulphate**( ) Hydroxyurea( ) Pain killers( ) Iron chelators( ) Don’t know10. Persons with sickle cell disease should be prescribed antibiotics and guided as to how to take them in all of the following situations EXCEPT FOR( ) Children under 5 years old**(answer) Pregnant women**( ) In dental procedures( ) In case of presumed infection( ) Don’t know11. In the case of teenagers with sickle cell disease, it is INCORRECT to state that( ) They will generally experience delayed growth and puberty( ) They are prone to develop self-concept, self-image and self-esteem disorders**(answer) Priapism in male teenagers is always related to sexual desire**( ) They may have academic and professional skills deficits( ) Don’t know12. As regards pregnancy and contraception in persons with sickle cell disease, it is CORRECT to state that( ) Urinary and respiratory tract infections are uncommon**(answer) Pregnancy is a potentially serious condition for woman and fetus**( ) Women with sickle cell disease must be advised against becoming pregnant ( ) Pre-natal care must be done at a primary healthcare clinic( ) Women with sickle cell disease must not be on injectable or oral contraceptives( ) Don’t know13. All of the following are recommended guidelines for preventing leg ulcers in persons with sickle cell disease EXCEPT FOR( ) Skin nourishing and cleansing( ) Use of adequate and comfortable footwear( ) Use of insect repellents and insect bite treatment**(answer) Mandatory use of compression stockings**( ) Do not know |