**Appendix 1:** DFConhecimento instrument

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| **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 know  2. 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 know  3. Which one is the Sickle Cell Anemia Genotype?  ( ) Hb SC  ( ) Hb AS  **(answer) Hb SS**  ( ) Hb SD  ( ) Hb AA  ( ) Don’t know  4. 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 know  5. 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 know  6. 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 know  8. 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 know  9. 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 know  10. 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 know  11. 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 know  12. 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 know  13. 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 |