***Table 1: Demographic characteristics of patients***

|  |  |  |
| --- | --- | --- |
| **Characteristic** | **Frequency** | **Percentage** |
| Sex  |  |  |
|  Female | 1,483 | 47.1 |
|  Male | 1,665 | 52.9 |
| Age category  |  |  |
|  0 – 4+ years | 869 | 27.6 |
|  5-9+ years | 915 | 29.1 |
|  10-14+ years | 821 | 26.1 |
|  15-17+ years | 258 | 8.2 |
|  >=18 years | 285 | 9.1 |
| SCD Phenotype |  |  |
|  SCD-SS | 2,116 | 67.2 |
|  SCD-SC | 1,023 | 32.5 |
|  Sβ+Thal  | 9 | 0.3 |

***Table 2: SCD-related diagnosis***

|  |  |
| --- | --- |
| **Diagnosis** | **Incidence Rate per 10,000 person months (95% CI)** |
| **SCD-SS** | **SCD-SC** | **SCD S Beta-plus thalassemia** |
|  **(Total Person months = 18, 328.14)** | **(Total Person months = 5, 662.55)** |  **(Total Person months = 125.52)** |
| Sickle cell painful event | 141.86 (125.73, 160.06) | 203.09 (169.48, 243.36) | 159.34 (40.29, 630.08) |
| Severe anemia secondary to hyper hemolysis | 32.74 (25.43, 42.15) | 14.23 (7.07, 28.24) | 0 |
| Acute malaria | 17.46 (12.35, 24.68) | 12.36 (5.90, 25.92) | 0 |
| Cerebrovascular accident (CVA) | 12.55 (8.34, 18.88) | 3.53 (0.88, 14.12) | 0 |
| Acute chest syndrome (ACS) | 7.64 (4.53, 12.90) | 5.30 (1.71, 16.42) | 0 |
| Avascular necrosis (AVN) | 7.09 (4.12, 12.21) | 21.19 (12.04, 37.29) | 0 |
| Dactylitis | 5.46 (2.94, 10.14) | 3.53 (0.88, 14.12) | 79.67 (11.31, 561.17) |
| Sepsis | 3.82 (1.82, 8.01) | 7.06 (2.65, 18.82) | 0 |
| Priapism | 2.73 (1.14, 6.56) | 1.77 (0.25, 12.54) | 0 |
| Osteomyelitis | 2.18 (0.82, 5.81) | 1.77 (0.25, 12.54) | 0 |

**SCD-SS – Sickle Cell Disease SS; SCD –SC – Sickle Cell Disease SC**

**Table 3:** **Definition of SCD terms**

|  |  |
| --- | --- |
| **Name** | **Definition** |
| SCD-SS | A variant of SCD due to homozygosity of the E6V mutation, amino acid substitution of valine for glutamic acid in the sixth position of the beta globin chain, resulting in the production of hemoglobin S from both alleles. |
| **SCD- S beta-zero thalassemia** | A form of sickle cell thalassemia characterized by the absence of hemoglobin A. Patients usually have severe anemia identical to that seen in sickle cell disease. |
| **SCD S Beta-plus thalassemia** | A mild form of sickle cell thalassemia characterized by the presence of hemoglobin S and a reduced amount of hemoglobin A in the red blood cells. It is characterized by the presence of small red blood cells and mild anemia.  |
| **SCD- SC** | A type of sickle cell disease characterized by the presence of both hemoglobin S and hemoglobin C. It is similar to, but less severe than SCD-SS. |
| **Steady State** | That period when the patient with sickle cell disease is not experiencing infections, pain, or other acute disease complications |
| **Sickle cell painful event** | Pain lasting at least two hours that requires an unscheduled emergency room visit or hospitalization or that disrupts daily activities |
| **Acute Chest Syndrome (ACS)** | A lung disease that involves a vasoocclusive crisis of the pulmonary vasculature seen in patients with sickle cell disease.  |
| **Non-specific acute lower respiratory tract episodes** | Includes acute respiratory episodes with lower respiratory tract signs that do not meet the criteria for other diagnoses. May include episodes which would have been diagnosed as ACS were radiographic facilities available. |
| **Acute anemia** | A “generic” term for a sudden drop (often defined as 20% or more) in Hb level beyond the baseline and divided into three common pathophysiologic types: “Acute splenic sequestration”, “Transient erythroid aplasia” (most commonly due to parvovirus B19 infection, and “Acute hemolysis” of various causes,) |
| **Hyperhaemolysis** | Significant change in blood picture characterized by a precipitous fall in the hemoglobin level associated with jaundice, marked reticulocytosis, and polychromasia on the blood smear, increased unconjugated hyperbilirubinemia, and increased urobilinogen content in urine above the steady state level for each individual patient. |
| **Malaria** | An individual with malaria-related symptoms (fever -axillary temperature ≥ 37.5°C, chills, severe malaise, headache or vomiting) at the time of examination or 1–2 days prior to the examination in the presence of a *Plasmodium* positive blood smear or a positive Malaria Rapid Diagnostic Test |
| **Cerebrovascular accident (CVA)** | Characterized by sudden loss of neurological function due to brain ischemia or intracranial hemorrhages. Presents with sudden onset of weakness, aphasia and sometimes seizures or coma and results in adverse motor and cognitive sequelae |
| **Avascular necrosis (AVN)** | Also known as aseptic necrosis, osteonecrosis or ischemic necrosis is bone death due to compromised blood supply. The hip joint is the most common site of AVN. |
| **Dactylitis** | A severe acute inflammatory response affecting the hands and feet of individuals with sickle cell disease. It is caused by vaso-occlusive episodes leading to ischemia and finally infarction of the distal portions of the extremities. Clinical signs of pain, swelling and tenderness of digits usually begin in early childhood and may be the initial manifestations of sickle cell anemia |
| **Sepsis** | The body’s severe inflammatory response to infection and mostly presents with fever. Infections that lead to sepsis most often start in the lung, urinary tract, skin or gastrointestinal tract. |
| **Priapism** | A sustained unwanted painful erection lasting 4 or more hours and not associated with sexual activity |
| **Osteomyelitis** | An inflammatory process accompanied by bone destruction and caused by an infecting microorganism |

Table 4: List of database parameters

| **Field Label** | **Choices, Calculations, OR Slider Labels** |
| --- | --- |
| Record ID |   |
| Enrollment pathway | 0, Clinic | 1, Mass registration | 2, Home visit |
| Name of person signing consent |   |
| Relationship to participant | 0, Biological Mother | 1, Biological Father | 2, Grandparent | 3, Aunt/Uncle | 4, Self | 5, Other |
| Other relation |   |
| Name of Participant/Child |   |
| Consent indication | 0, Thumbprint|1, Signature |
| Consent Date |   |
| Person Obtaining Consent |   |
| OP Unit Number |   |
| Patient ID/ LHIMS Number |   |
| Visit Date (dd/mm/yyyy) |   |
| Hospital Name | 0, KATH | 1, KNUST | 3, Kumasi South |
| Diagnosis Pathway | 0, Non-NSP|1, NSP |
| NSP Number |   |
| First Name |   |
| Middle Name |   |
| Last Name |   |
| Date of Birth (dd/mm/yyyy) |   |
| Date of Birth Correctness | 1, exact date | 2, exact day | 3, month and year | 4, year only | 5, unknown |
| Calculated age at recruitment (yr) | rounddown(datediff ([date\_of\_birth], [consent\_date], "y", "dmy"),0) |
| Calculated age at recruitment (mths) | rounddown(datediff ([date\_of\_birth], [consent\_date], "M", "dmy"),0) |
| Age at registration (months) {Participant less than 5 years} |   |
| Age at registration (completed years) {Participant more than 5 years} |   |
| Marital Status | 1, Single|2, Married|3, Other|4, Cohabiting|5, Divorced|6, Widowed |
| Sex | 1, Male | 0, Female |
| Occupation |   |
| Religion | 0, None|1, Christian|2, Muslim|3, Traditionalist|4, Other |
| Other religion |   |
| Educational Status | 0, None | 1, Basic | 2, JHS | 3, SHS/Vocational | 4, Tertiary |
| Region |   |
| Residence |   |
| Residential Status | 1, Rural | 2, Semi-urban | 3, Urban |
| Digital Address System |   |
| Telephone1 |   |
| Telephone2 |   |
| Year of Diagnosis |   |
| SCD Test Result (SS/SBThal/SC) | 1, SS | 2, SBThal | 3, SC | 4, S-beta thal zero (nought) | 5, S-beta thal plus |
| ABO Blood Group | 1, A+ | 2, A- | 3, B+ | 4, B- | 5, O+ | 6, O- | 7, AB+ | 8, AB- | 9, Unknown |
| Pneumococcal Vaccination up to date | 0, No|1, Yes|9, Unknown |
| Penicillin V(prophylaxis) | 1, Yes|0, No|3, NA|9, Unknown |
| Folic Acid |   |
| Using Hydroxyurea |   |
| Date of Initiation of Hydroxyurea Therapy |   |
| Anti Malaria Prophylaxis | 1, Yes|0, No|3, NA|9, Unknown |
| Visit date (dd/mm/yyyy) |   |
| Temperature |   |
| Weight (Kg) |   |
| Length/Height (cm) |   |
| Pulse Rate (bpm PULSE) |   |
| Respiratory Rate (cpm) |   |
| Oxygen Saturation (%) |   |
| Systolic Blood Pressure (mmHg SBP) |   |
| Diastolic blood Pressure (mmHg DBP) |   |
| Head Circumference (cm) |   |
| Reason for Visit | 0, Routine|1, Acute|2, Review|3, First visit |
| Admissions | 1, Yes|0, No |
| Admission details |   |
| Transfusion | 0, No|1, Yes|9, Not assessed |
| Units of Blood |   |
| Details of Transfusion |   |
| Nutrition | 0, Normal|1, Abnormal|9, Not assessed |
| Details of Nutrition |   |
| Growth | 0, Normal|1, Abnormal|9, Not assessed |
| Details of Growth |   |
| Development | 0, Normal|1, Abnormal|9, Not assessed |
| Details of Development |   |
| Menstral/Sexual | 0, Normal|1, Abnormal|9, Not assessed |
| Details Menstral/Sexual |   |
| Ferile Illness | 0, None|1, Present|2, Past|9, Not assessed |
| Details of Ferile illness |   |
| Pain | 0, None|1, Present|2, Past|9, Not assessed |
| Details of Pain |   |
| Enuresis | 0, None|1, Present|2, Past|9, Not assessed |
| Details of Enuresis |   |
| Priaprism | 0, None|1, Present|2, Past|9, Not assessed |
| Details of Priaprism |   |
| CNS | 0, None|1, Present|2, Past|9, Not assessed |
| Details of CNS  |   |
| Cough | 0, None|1, Present|2, Past|9, Not assessed |
| Details of Cough |   |
| Difficult in Breathing | 0, None|1, Present|2, Past|9, Not assessed |
| Details of Difficult in Breathing |   |
| Other Symptoms | 0, None|1, Present|2, Past|9, Not assessed |
| State the details of other symptoms |   |
| Are you Pregnant | 1, Yes|0, No|3, NA|9, Unknown |
| Date of EDD (dd/mm/yyyy) |   |
| Additional Interim History |   |
| Social Family Relations |   |
| School Attendance/Performance |   |
| Immunization Done | 1, DPT/dT|2, Polio|3, Hib|4, Hep|5, MMR|6, Pneumo |
| Skin | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal skin details |   |
| Pallor | 1, Yes|0, No |
| Head | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal head details |   |
| Ears | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal ear details |   |
| Anaemic | 1, Yes|0, No |
| Details of Anaemia |   |
| Jaundice | 0, Nil|2, Tinge|3, Moderate|4, Deep |
| Details of Jaundice |   |
| Eyes | 0, Normal|1, Abnormal|9, Not assessed |
| Other abnormal eye details |   |
| Fundi | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal fundi details |   |
| Nose | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal nose details |   |
| Mouth | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal mouth details |   |
| Teeth | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal teeth details |   |
| Throat | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal throat details |   |
| Neck | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal neck details |   |
| Nodes | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal nodes details |   |
| Chest | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal chest details |   |
| Lungs | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal lungs details |   |
| Heart | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal heart details |   |
| Abdomen | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal abdomen details |   |
| Liver | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal liver details |   |
| Spleen | 0, Normal|1, Abnormal|9, Not assessed |
| Spleen Palpable (size) |   |
| Other abnormal spleen details |   |
| Genitalia | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal genitalia details |   |
| Vertebrae | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal vertebrae details |   |
| Extremities | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal extremities details |   |
| Joints | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal joints details |   |
| Neurological | 0, Normal|1, Abnormal|9, Not assessed |
| Abnormal Neurological details |   |
| Lab test results | 0, None|1, FBC/CBC|2, Biochemistry|3, LFT|4, HB Electrophoresis|5, Urine RE|6, Blood Culture|7, Other|8, ESR|9, RFT|10, MPs |
| FBC test date |   |
| WBC (10^3uL) |   |
| RBC (10^6uL) |   |
| HB (g/dL) |   |
| MCV (fL) |   |
| MCH (pg) |   |
| MCHC (g/dl) |   |
| PLT (10^3ul) |   |
| RETICS % |   |
| ESR |   |
| Mps | 10, RDT Negative|11, RDT Positive|20, Microscopy negative|21, Microscopy Positive |
| LFT Date |   |
| AST (u/l) |   |
| ALT (u/L) |   |
| ALP (u/L) |   |
| GGT (u/L) |   |
| Albumin (g/L) |   |
| Bilirubin total (umol/L) |   |
| Total protein (g/L) |   |
| Globulin (g/L) |   |
| Serum Electrolyte date |   |
| Sodium (Na) mmol/L |   |
| Potassium (K) mmol/L |   |
| Chloride (Cl) mmol/L |   |
| Total CO2 mmol/L |   |
| RFT Date |   |
| Urea (mmol/L) |   |
| Creatinine (mmol/L) |   |
| BUN to Creatinine Ratio |   |
| Other tests results |   |
| HB Electrophoresis date |   |
| Hb Genotype |   |
| Appearance |   |
| Colour |   |
| pH |   |
| Protein | 0, Negative|1, Positive |
| Glucose | 0, Negative|1, Positive |
| Ketone | 0, Negative|1, Positive |
| Blood | 0, Negative|1, Positive |
| Bilirubin | 0, Negative|1, Positive |
| Nitrite | 0, Negative|1, Positive |
| Urobilinogen | 0, Normal|1, Abnormal |
| Specific gravity |   |
| Pus cells (hpf) |   |
| Red blood cells (/hpf) |   |
| Epithelial Cells (/hpf) |   |
| RBCs | 0, Not seen|1, Present |
| Crystals | 0, Not seen|1, Present |
| Casts | 0, Not seen|1, Present |
| Yeast cells | 0, Not seen|1, Present |
| T. Vaginals | 0, Not seen|1, Present |
| S. Haematobium | 0, Not seen|1, Present |
| Spermatozoa | 0, Not seen|1, Present |
| Clue Cells | 0, Not seen|1, Present |
| Others | 0, Not seen|1, Present |
| Other Urine RE findings |   |
| Imaging Results Available | 0, None|1, X-ray|2, TCD|3, Ultrasound|4, CT|5, MRI |
| X-ray Date |   |
| Xray results |   |
| TCD date |   |
| TCD result MCA\_left |   |
| TCD result MCA\_Right |   |
| TCD result ICA\_left |   |
| TCD result ICA\_Right |   |
| TCD remarks |   |
| Ultrasound date |   |
| Ultrasound results |   |
| CT Date |   |
| CT results |   |
| MRI Date |   |
| MRI results |   |
| Overall assessment |   |
| Issues and Problems |   |
| Tests ordered | 1, FBC | 2, Electrolyte | 3, X-ray | 4, Ultrasound | 5, TCD | 6, LFT | 7, RFT | 8, Retics | 9, MPs | 10, BUE Cr | 11, Blood Culture | 12, Urine RE |
| Routine Medications | 1, Penicilin V 125mg bd|2, Pennicillin V 250mg bg|3, Folic acid 5mg dly|4, Folic Acid 10mg dly|5, HU|6, Haematinics|7, Analgesics |
| Medications Prescribed |   |
| Other recommendations for issues and problems |   |
| Referrals |   |
| Next Routine Visit (dd/mm/yyyy) |   |
| Next Visit Correctness | 1, exact day, month and year | 2, date unknown |
| Attending Doctor |   |
| Today's Education Topics |   |