NEWSLETTER

SICKLE CELL DISORDER REGISTRY NIGERIA (SCDRN)

April, 2023

Data gathering is critical to providing adequate and appropriate treatments and care for persons living with Sickle Cell Disease

Contents

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SICKLE CELL DISEASE REGISTRY NIGERIA

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BACKGROUND

Nigeria has the highest burden of sickle cell disease in the world. An estimated 150,000 babies are born every year with sickle cell anaemia (Hb SS).

Sickle cell disease (SCD) is the most common clinically significant haemoglobinopathy, characterised by painful episodes, anaemia, high risk of infection, and other acute and chronic complications.

SCD is a major genetic condition in Nigeria. The country has by far the largest burden of the disorder anywhere in the world. Over 40 million Nigerians are carriers of the sickle cell gene and an estimated 150,000 babies are born every year with sickle cell anaemia (Hb SS). Sadly, about 100,000 of these babies do not live to celebrate their fifth birthday; they die mostly from ignorance and lack of access to proper diagnosis and care.

100,000 children die before five years mostly from lack of knowledge and access to proper diagnosis and care

History of the Sickle Cell Disorder Registry Nigeria (SCDRN)

Over the years, statistics about Sickle Cell Disease Management in Nigeria have been based on estimates. This has impacted negatively on policymaking and the quality of care available for the effective management of the disorder in Nigeria. In a bid to change the narrative and make data readily available to healthcare professionals as well as policymakers, the Sickle Cell Foundation Nigeria 2018 signed a Memorandum of Understanding with PointCare Health Initiative (Rhieos-Ventures) based in Netherlands, to set up the multi-centre SCD registry for persons with (SCD). The initiative which commenced with a mandate to ultimately collect data from all locations across Nigeria where care and treatment for persons living with SCD is obtained began at - the Lagos University Teaching Hospital Idi-Araba Paediatric and Adult clinics. These are still the primary data collection centres till date.

What is SCDRN?

The SCDRN is built on an online platform called - Research Electronic Data Capture (REDCap). It is a simple, efficient and secure web application that is utilised to capture both online and offline routine data at our various clinics. The registry is a powerful tool to observe the course of disease; understand variations in treatment and outcomes, examine factors that influence prognosis and quality of life, describe care patterns, including appropriateness of care and disparities in the delivery of care.

What Information is collected and how is data used?

Data collected include; bio-demographics, past medical history, clinical signs, and symptoms, treatment, disease condition/complications, etc. Participants on the registry are enrolled after giving their consent to be included in the registry. Subsequently, are captured at each clinic visit. Data from the registry would be used to inform evidence-based management of SCD, improve research as well as give policy makers - information and tools with which to guide policy formulation and laws that would be beneficial to the sickle cell community.

The registry became operational in 2019, but activities were truncated due to the outbreak of COVID-19 in early 2020. Happily, work in the registry resumed in February 2022, with active data capture again.

How is the data captured?

The registry is designed as a multi-centre registry where outpatient information is collected at each clinic visit. At the moment, there are two centres/clinics in the Lagos University Teaching Hospital (LUTH) - the Paediatric clinic and Adult haematology clinic.

Objectives

The objectives of the SCD Registry are inter alia - to:

1. Collect accurate clinical and patient-reported data on persons with SCD as per registry protocol.
2. Promote clinical research into interventions.
3. Support the management and quality of care of persons with SCD.
4. Foster health promotion, health education, and disease prevention programmes
5. Enhance the capacity of health care providers.

ACHIEVEMENTS OF THE REGISTRY

Over the last 12 months of active data capture at the Paediatric and Adult clinics in Lagos University Teaching Hospital (LUTH), the registry has recorded phenomenal success in the number of persons who are enrolled.

SICKLE CELL DISORDER

ACHIEVEMENTS OF THE SICKLE CELL DISEASE REGISTRY NIGERIA

1. Over 100% increase in data captured between 2019 and 2022.
2. Presentation of the registry outcomes at a conference.
3. The Registry engaged a number of key personnel – Data Collection Officers and Research/database officer.

DATA CAPTURED

Over 100% increase in data captured between 2019 to 2020 and 2022.

PRESENTATION

Presentation of the registry outcome.

STAFF

Data Collection Officers as well as a Research/database officer engaged.

Registry data as 31st March 2023

- Total number of subjects in registry: 987
- Total number with >2 SCD in family: 28
- Average no. of persons in household: 6
- Age of oldest Participant in registry: 67
- Age of youngest Participant: 3 Months
- Average Age at Diagnosis of SCD: 3 Years Old
- Mean Follow-up Visits: 2

IMPLICATIONS OF RESULTS FROM THE REGISTRY

We currently have 987 patients in our Registry. Key research questions that have been addressed by the registry include the following:

- Average age at diagnosis of sickle cell is 3 years; this may be an indication that we are not diagnosing sickle cell disease early enough, so that prompt treatment can be instituted to prevent morbidity and mortality.

- Oldest patient in the registry is aged 67 years – indicating that persons with sickle cell live into the 7th decade of life.

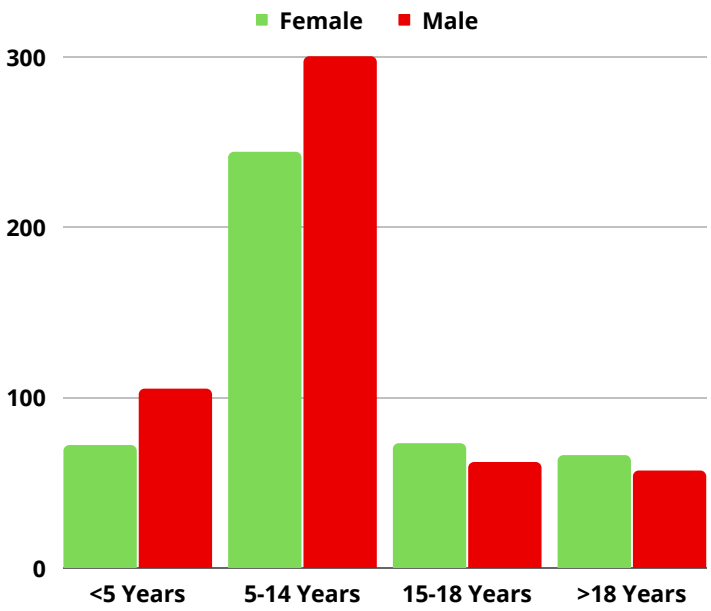
- Mean Follow-up visits is 2 - an indication that there may be need for a more robust protocol for follow-up of patients.

- Number of families with 2 or more persons living with sickle cell is 28 – an indication of high burden on these families.

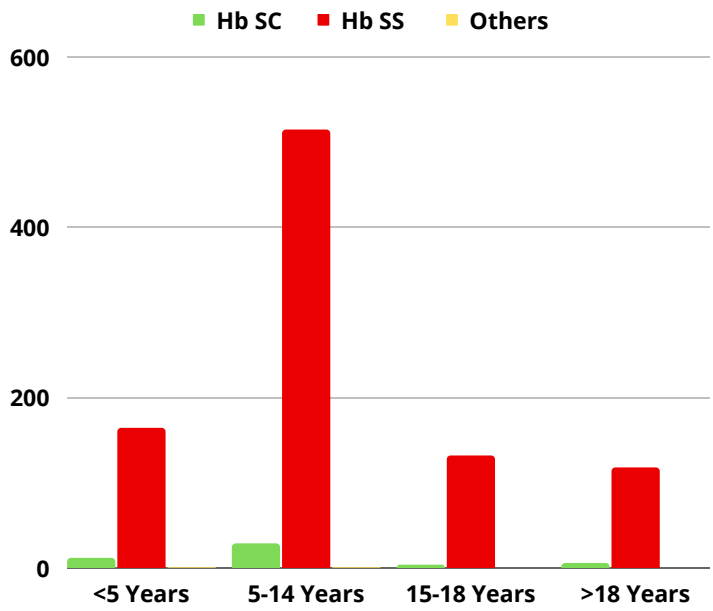
- Average number of people in households is 6 - indicative of heavy socio-economic burden on families.

Bar charts showing Distribution of Age, Sex, Haemoglobin Phenotype, Transfusion History, Hydroxyurea Therapy (Numbers)

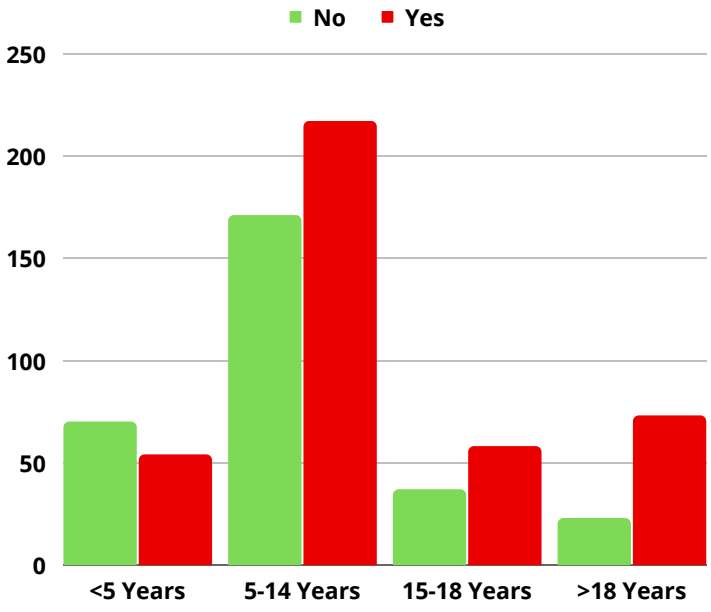
Sex



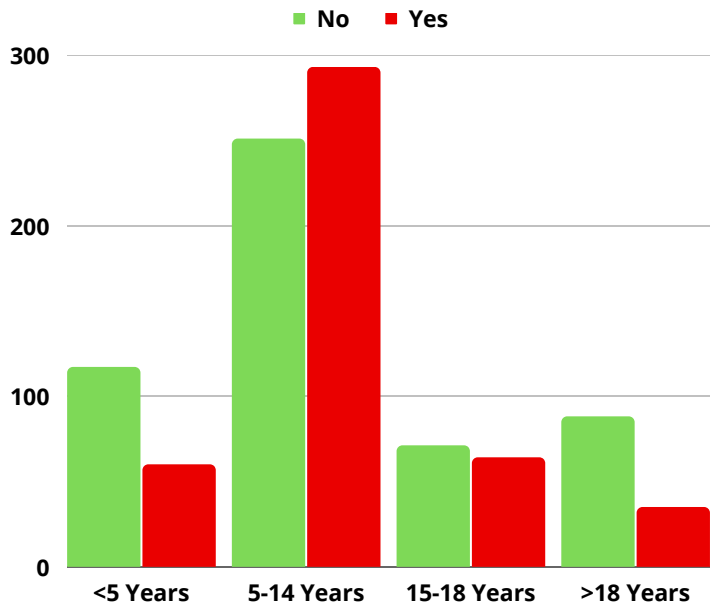
Haemoglobin phenotype



Transfusion history

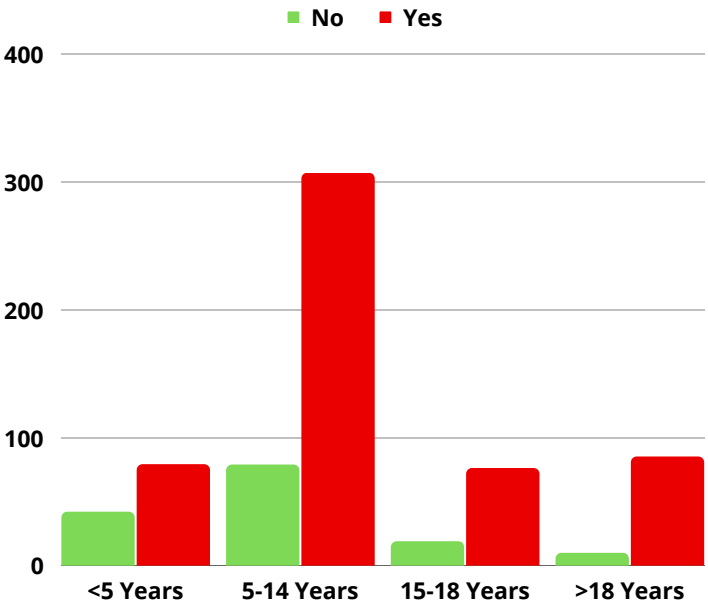


Hydroxyurea therapy

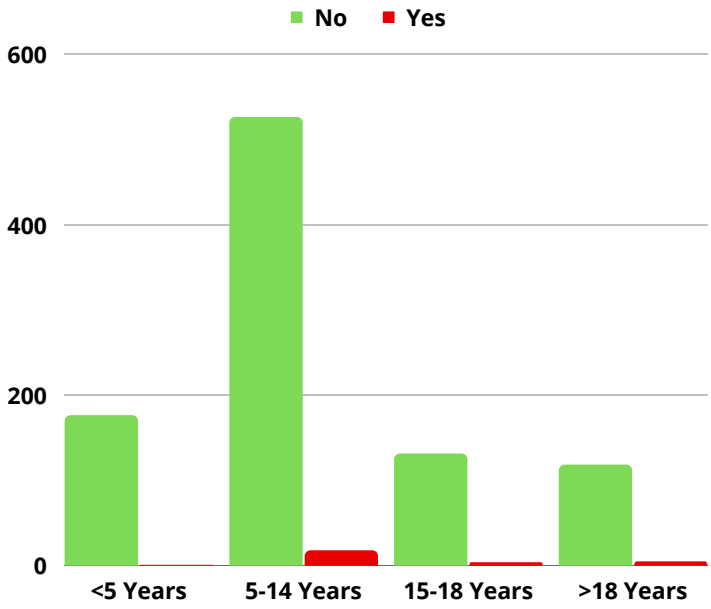


Bar charts showing distribution of Hospitalisation History, Stroke, Acute Chest Syndrome, History of Heart Failure (Numbers)

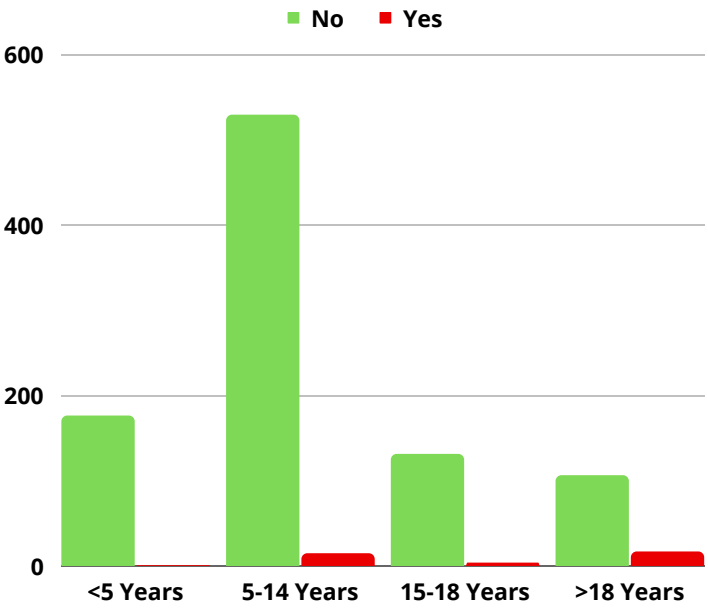
Hospitalization history



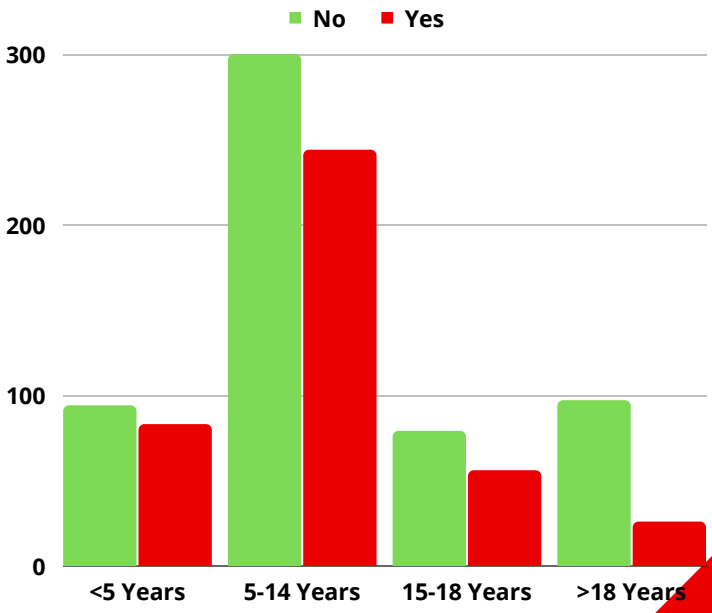
Stroke



Acute chest syndrome

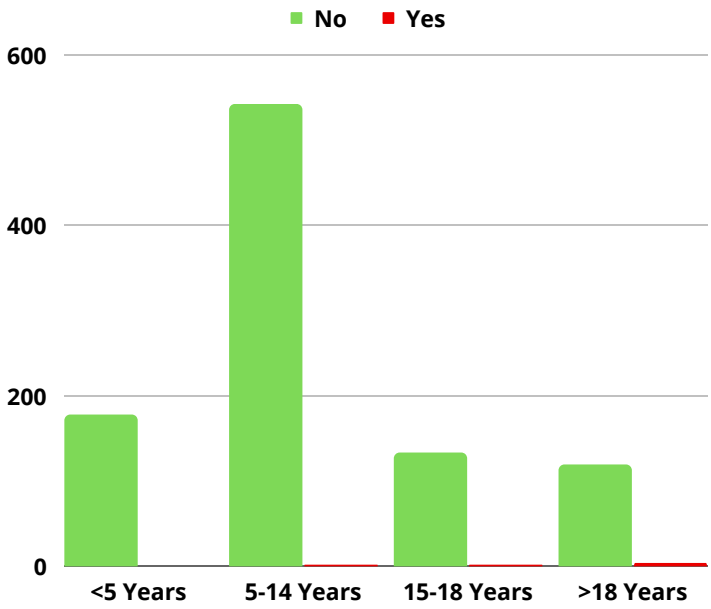


History of heart failure

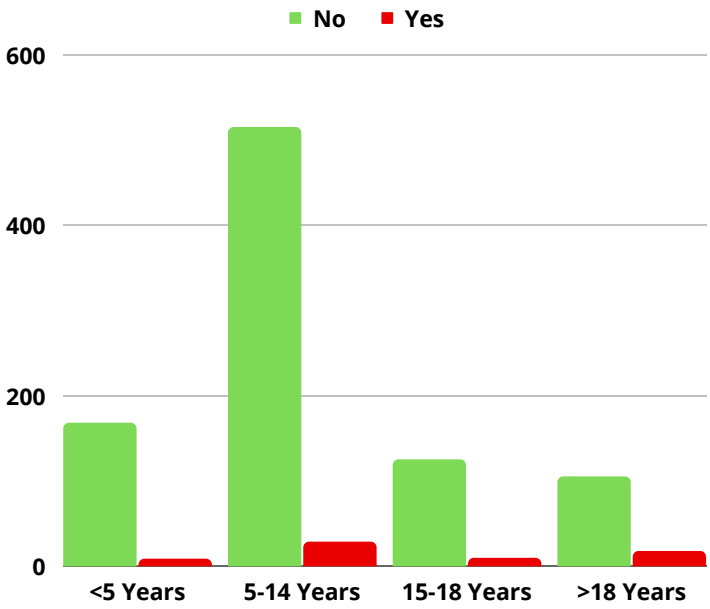


Bar Charts showing the distribution of Kidney Dysfunction, Priapism, Trans-Cranial Doppler (TCD) scan for stroke (Numbers)

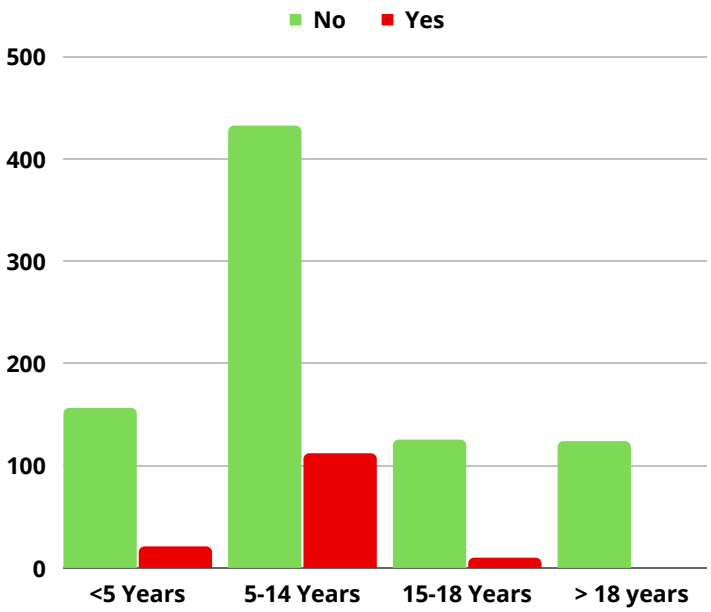
Kidney dysfunction



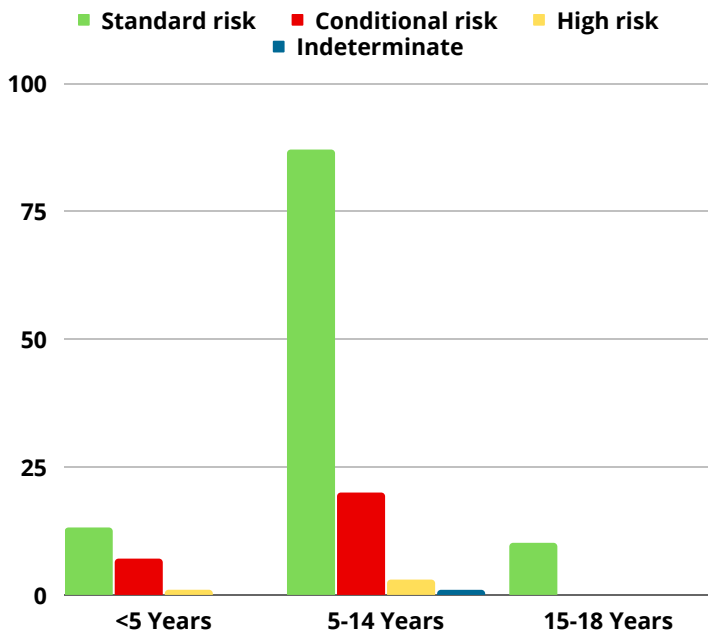
Priapism (Males Only)



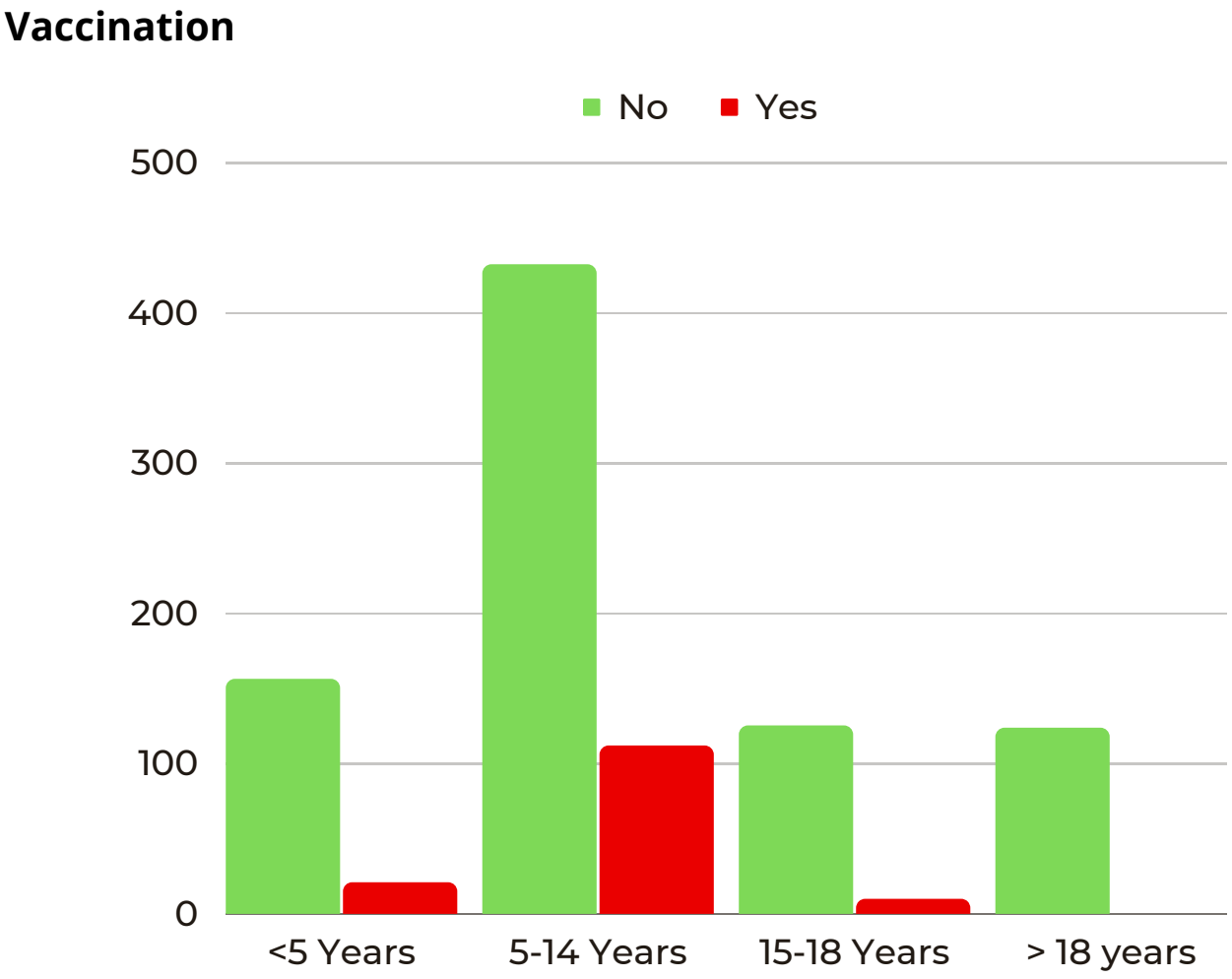
Those who have done TCD



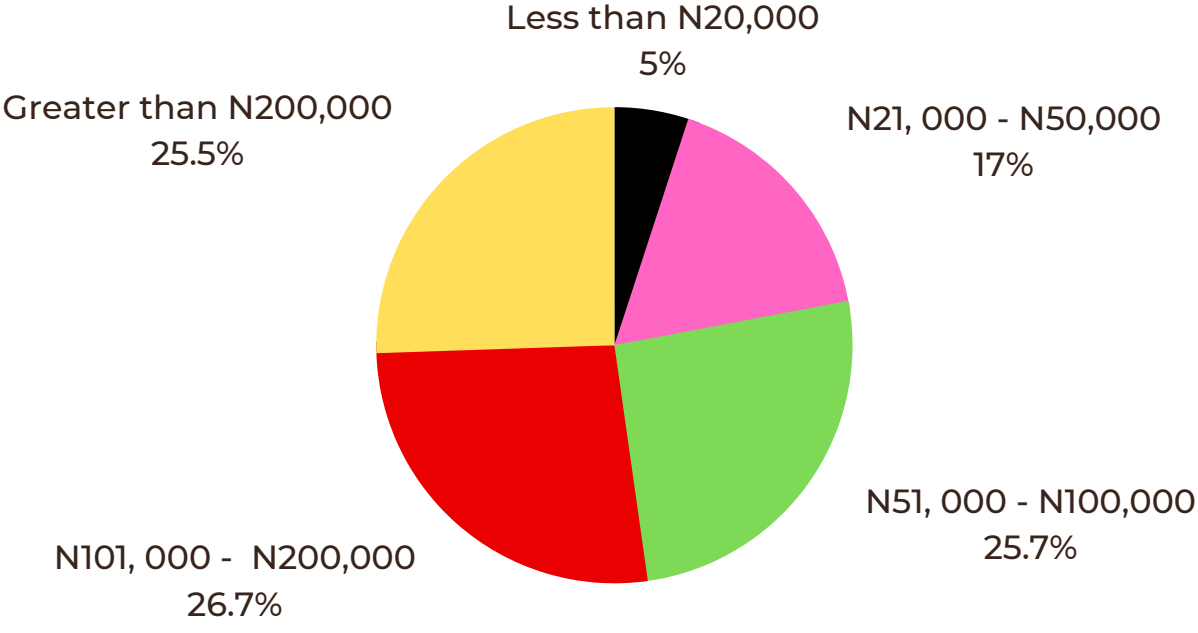
Trans-Cranial Doppler Test result



Bar Chart showing the distribution of vaccination (Numbers)



Pie Chart showing Estimated Total Family Income (Per Month)



WAY FORWARD FOR THE REGISTRY IN 2023

In 2023, among other ongoing activities, the plan is to:

1. Build on the gains of the last 12 months.
2. Expand the registry to other facilities within and outside Lagos state.
3. Attract more funding and deepen collaborations with existing and new organisations to implement plans.
4. Report/disseminate the registry statistics and outcomes data quarterly.
5. Publish peer-reviewed articles based on the registry outcomes data

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