Newborn screening for sickle cell disease in Africa

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I declare no competing interests

For more on the **Consortium on Newborn Screening in Africa** see https://www.hematology.org/global-initiatives/consortium-on-newborn-screening-in-africa

For the **1986 study** see *N Engl J Med* 1986; **314:** 1593–99

For more on the **ARISE project** see https://www.ariseinitiative.org/

For more on **screening in Nigeria** see **Articles** *Lancet Haematol* 2020; **7:** e534–40

On April 21, 2021, newborn screening for sickle cell disease was launched at Arthur Davison Children's Hospital in Ndola, Zambia. The first newborn screening of sickle cell disease and early therapeutic intervention programme within Zambia's existing framework of the Expanded Programme on Immunization and HIV early infant diagnosis is part of the ongoing American Society of Hematology's Consortium on Newborn Screening in Africa (CONSA). Screening will be done at three hospitals in Zambia to create a registry of demographic, medical, and laboratory information of children diagnosed with sickle cell disease.

Launched in 2016 to address the global burden of sickle cell disease, CONSA shows the value of newborn screening and how it can be implemented in diverse settings throughout sub-Saharan Africa. "More than 300 000 babies are born with sickle cell disease in sub-Saharan Africa every year. Yet, the vast majority do not live to the age of 5", says Kwaku Ohene-Frempong, President of the Sickle Cell Foundation of Ghana and CONSA national coordinator. "This is due mainly to three largely preventable causes: invasive pneumococcal infection, acute malaria, and acute splenic sequestration, all of which can be associated with high rates of mortality after a short illness", he told *The Lancet Haematology*.

Early diagnosis of sickle cell disease followed by twice daily oral penicillin prophylaxis greatly reduces pneumococcal bacteraemia; malarial chemoprophylaxis or insecticide-treated bed nets help to reduce the incidence of and mortality from acute malaria infection; and parental and caregiver training on palpation of the spleen allows early diagnosis of splenomegaly due to acute splenic sequestration and prompt medical consultation for life-saving intervention, Ohene-Frempong explains. The life-saving value of newborn screening for sickle cell disease followed by penicillin prophylaxis was shown in the USA in 1986. "These inexpensive interventions reduced the mortality of babies with sickle cell disease in Kumasi, Ghana, to 4·5% in the first 10 years of life in the pilot newborn screening programme", Ohene-Frempong says.

CONSA's goal is to introduce standard-of-care practices for screening and early-intervention therapies at participating institutions, screening 10 000–16 000 babies per year in Kenya, Ghana, Liberia, Nigeria, Uganda, Tanzania, and Zambia, and providing clinical follow-up for babies who test positive for the disease, says Enrico Novelli, CONSA co-chair. Haematologists and public health officials in CONSA mobilise networks of screening laboratories, sickle cell disease or paediatric haematology clinics, teaching hospitals, universities, and satellite clinics to screen babies and provide clinical services. Screenings have started in Ghana, Nigeria, Uganda, and Zambia; Kenya, Liberia, and Tanzania will begin screening in the next few months, Novelli says.

In Nigeria, the first babies identified on the CONSA programme were in the Angwan Dodo Primary Health Care Centre and in the University of Abuja Teaching Hospital in December, 2020. By mid-April, 2021, three more babies were confirmed to have sickle cell disease in the CONSA Abuja Network. In the Kaduna Network, the first two babies were screened for sickle cell disease at the special care baby unit of Ahmadu Bello University Teaching Hospital Shika-Zaria, Kaduna, in April. "This African Research and Innovative Initiative for Sickle Cell Education (also known as ARISE) project is one of many concerted efforts by different NGOs to carry out newborn screening in the country" says Nnodu Obiageli, Professor of Haematology and Blood Transfusion, University of Abuja, Nigeria.

The Nigerian government at the federal level has established comprehensive newborn screening centres in each geopolitical zone of the country since 2012, she says, adding "some of the centres have been carrying out newborn screening, although not all are active."

Newborn screening of sickle cell disease in Africa is gaining attention, Obiageli says, but "new programmes have to become properly established before we can describe it as expanding". "Lack of technical expertise and trained manpower, and the need to source all reagents and equipment from outside the continent" hampers progress, she says.

The uptake of newborn screening for sickle cell disease in Africa has been slow. No African country has committed to universal screening as they have to childhood immunisations, Ohene-Frempong says. Few countries have a newborn screening programme, and only a small percentage of babies born annually in Africa have been screened for the disease, he says.

Although CONSA has been a catalyst for newborn screening through provision of infrastructure, reagents for the screening assays, and training of health-care workers to implement screening in the consortium countries, each country must have budgetary allocation to support newborn screening in the long term, Obiageli notes.

"Our goal is long-term sustainability for the programme," says Novelli. "We look forward to engaging sickle cell disease advocates, local governments, international organisations, and industry partners as we see the expansion of newborn screening for sickle cell disease in sub-Saharan Africa."

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