TRANSCRANIAL DOPPLER SCREENING PROGRAMME IN CHILDREN AND ADOLESCENTS WITH SICKLE CELL DISEASE: EXPERIENCE IN A DISTRICT GENERAL HOSPITAL IN THE UK

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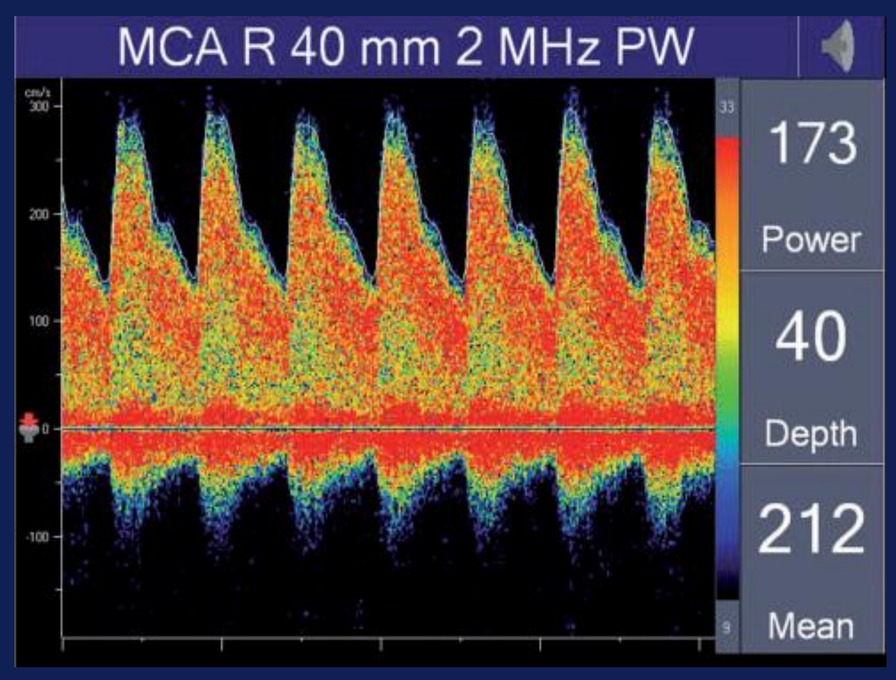
BACKGROUND

Sickle cell disease (SCD) is the one of the most common cause of stroke in the young worldwide. Without intervention, 11% of children with SCD are expected to have a stroke prior to the age of 20 years; nevertheless, the majority of these strokes are now considered preventable[1-3]. High cerebral blood velocities are associated with an increased risk of stroke, which can be assessed using transcranial Doppler (TCD)[4]. The Stroke Prevention Trial in Sickle Cell Anemia (STOP) demonstrated that stroke risk was reduced by 92% in children receiving chronic blood transfusions after detection of high blood velocities by TCD screening compared with standard care[5]. Annual TCD screening in children and adolescents with SCD between 2 and 16 years of age is now a standard of care[6,7]. Following this protocol, a sample of children screened for the first time would be expected to have a distribution of 70% normal, 15% conditional, 10% abnormal, and 5% inadequate TCD scans[8]. However, across the world, screening is inadequate, with rates between 22-44% of eligible children in the Unites States[9]; there are no European data on feasibility or adherence to the screening program.



Example of MCA TCD tracing in the "High risk range" obtained from a 7-year-old female HbSS patient

AIM

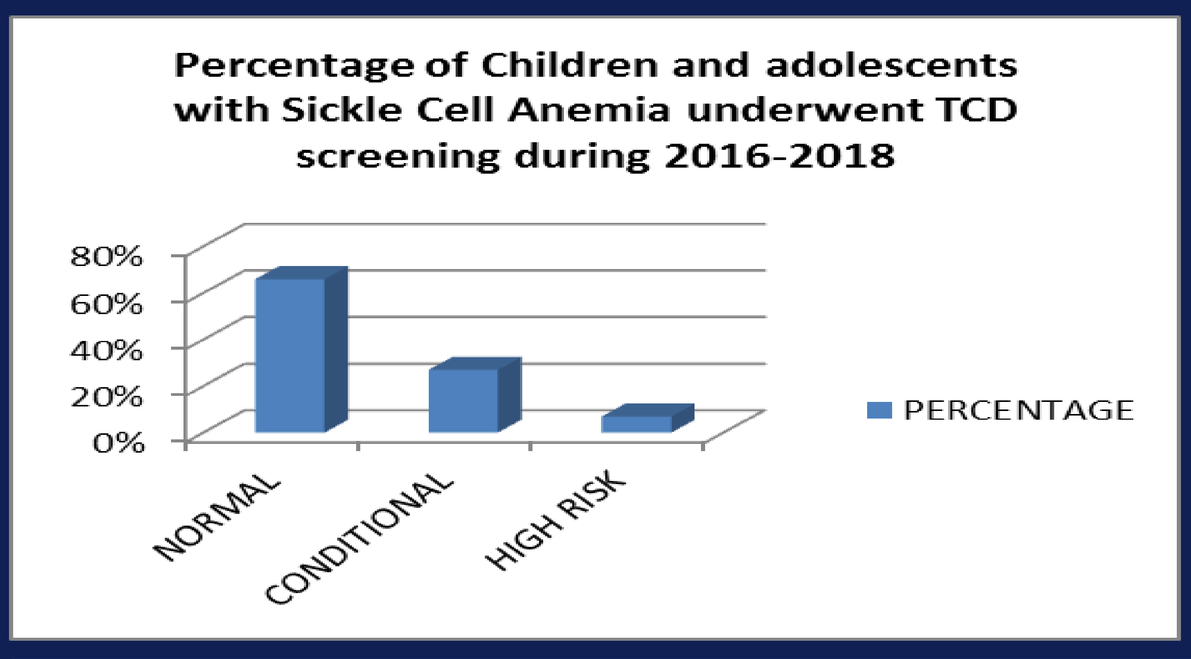
We aimed to investigate the feasibility of TCD screening program according to the STOP criteria, and screening rates, in a district general hospital (DGH) in the UK.

METHODS

Eligible children underwent local TCD screening as a tertiary paediatric support program from Oxford University Hospital. A dedicated specialist nurse liaised with children and families through home and school visits, facilitating attendance. Patients' clinical details, TCD time-averaged mean of the maximum velocity (TAMMV) and screening outcomes were recorded at each visit.

RESULTS

All 44 eligible SCD children attended the screening program (100% rate) between 2016 and 2018. Children with "time averaged mean of the maximum velocities" (TAMMV) < 170 cm/s in cerebral arteries were classified as "normal"; between 170-199 cm/s "conditional", > 200 cm/s "high risk", as per the STOP protocol.
Following the STOP trial recommendations, we observed among children screened a distribution of 66% normal, 27% conditional and 7% high risk. No patients were classified as inadequate. (Table)



2016-2018			
	NORMAL	CONDITIONAL	HIGH RISK
PATIENTS NUMBER	29	12	3
MEAN AGE 1st TCD (SD)	8.3 (5.1)	6.7 (3.5)	5.3 (2.1)
MEAN TAMMV (SD)	106.9 (26.5)	175.6 (5.2)	201 (1.7)
MEAN TCD/year (SD)	1 (0.1)	1.3 (0.25)	2.2 (0.3)
PERCENTAGE	66%	27%	7%

CONCLUSIONS

TCD screening for SCD is feasible at a UK DGH, with risk stratification close to the STOP trial recommendations. Specialist nurses working in the community have a critical role in promoting compliance to the program.

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