ICMR - Centre for Research, Management and Control of Haemoglobinopathies, Chandrapur

A comprehensive longitudinal cohort study for organ damage among sickle cell disease patients

Objectives

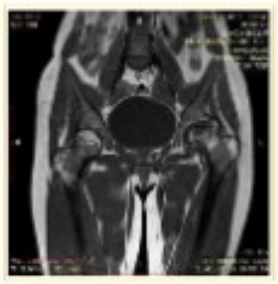
- To understand spectrum of chronic organ damage at different age groups
- To detect early organ dysfunction by conducting follow-ups
- To correlate effect of α-thalassemia and γ-gene modifiers
- To intervene patients with organ damage by Hydroxyurea therapy

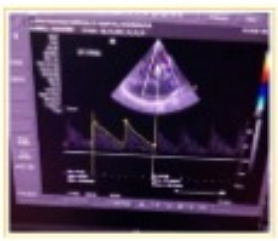
Materials and methods:

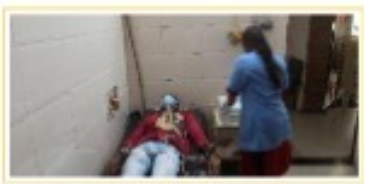
Clinical investigations: X-ray, MRI, Spirometry Transcranial doppler, Ultrasonography.

Laboratory investigations: Complete blood count, High performance liquid chromatography, Liver function test, Kidney function test.

Molecular investigations: Presence of sickle cell, β/α-thalassemia mutation, γ-globin gene mutation.



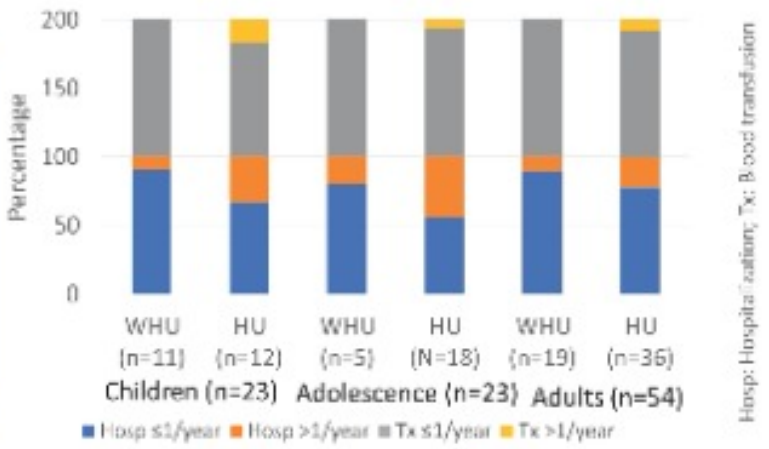




Results:

- 100 sickle cell disease (SCD) patients (45 Males: 55 Females) underwent clinical and laboratory investigations.
- SCD patients: Children (6-12 years): 23; Adolescents (13-17 years): 23; Adults (18-52 years): 54.
- 65% of patients were on hydroxyurea (HU) therapy. Majority of the patients on HU therapy had severe disease phenotype.
- The mean dosage of drug hydroxyurea patients receiving was 11.9 mg/kg/day.

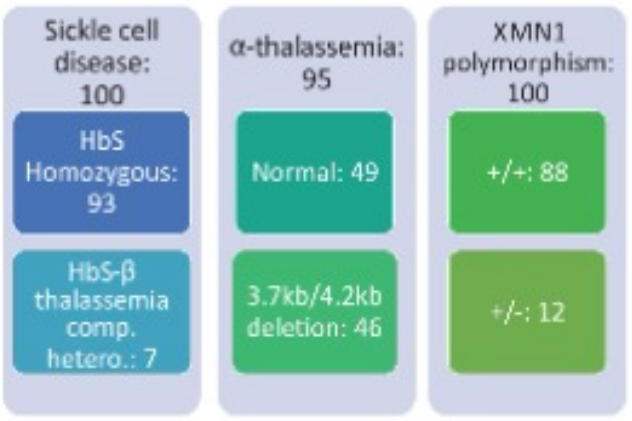
Clinical history and hydroxyurea status (Retrospective)



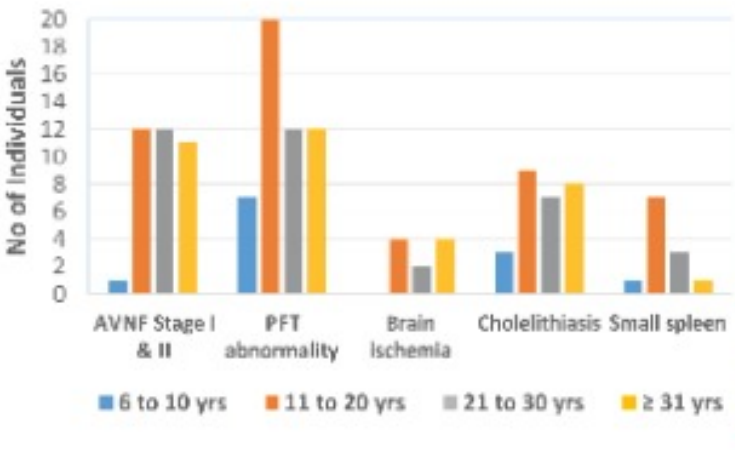
Hematological parameters

SCD patient	HU status	Hb (g/dL)	HCT (%)	MCV (fL)	HbF (%)	HbS (%)
Children	WHU (n=10)	8.2±1.5	25.8±3.7	79.3±9.1	21.3±5.9	71.2±8.0
	HU (n=12)	9.7±0.7	29.2±1.7	89.8±7.1	24.1±4.0	67.5±7.4
Adolescents	WHU (n=5)	8.5±1.3	25.9±3.8	83.3±4.1	17.9±3.5	74.1±6.9
	HU (n=15)	9.9±1.5	29.5±4.1	91.8±9.4	24.3±4.9	70.2±5.2
Adults	WHU (n=18)	9.3±1.5	28.5±4.6	82.3±8.9	19.0±7.5	75.3±8.1
	HU (n=35)	9.9±1.7	29.9±4.8	89.8±10	21.5±6.7	73.1±6.8

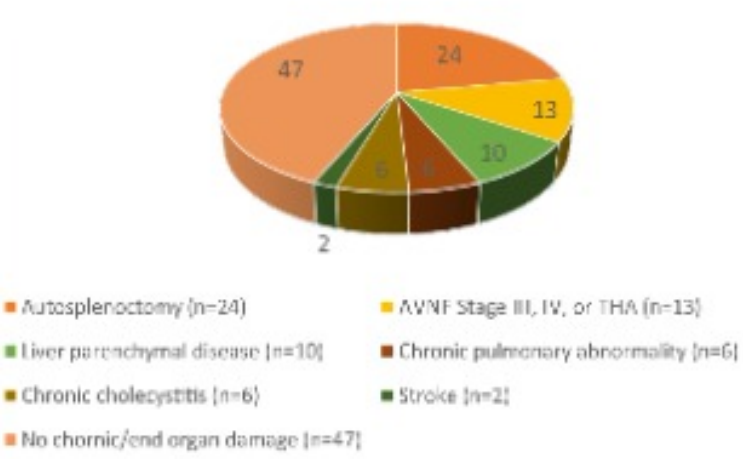
Molecular investigations



Early organ dysfunction in different SCD age groups



Chronic organ damage in 100 SCD patients



Conclusion:

- 53% SCD patients showed chronic organ damage.
- Early organ dysfunction commences in adolescent groups.

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