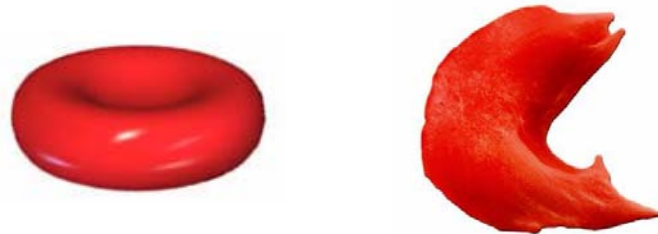


# Sickle cell disease in Cameroonian children:

The cause of severe cognitive deficits



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➤ Eagle Fondation

Nicolas Ruffieux  
Geneva Health Forum  
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# [ Plan ]

## 1. Sickle cell disease

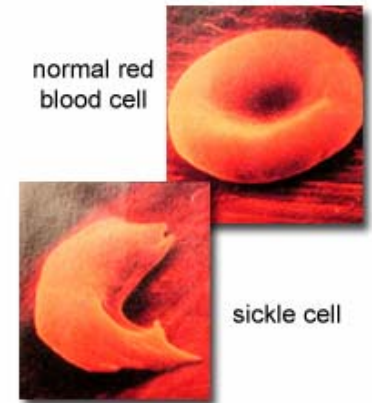
- Symptoms and complications
- Stroke
- Silent cerebral infarcts (SCI)
  - Cognitive repercussions of SCI
  - Study on SCI

## 2. The current project

- Objectives
- Method
- Results
- Conclusions

# Sickle cell disease (SCD)

- Inherited blood disorder affecting red blood cells
  - Increased adhesiveness to vascular endothelium → vaso-occlusion
- Most frequent in sub-Saharan countries
- Prevalence in Cameroon: 2-3% (Ministry of Public Health, Cameroon)
- Major complications : infections and stroke



Distribution of Sickle Cell Gene

# [ Stroke ]

- Most disabling complication
- At least 6.7% of SCD children in Yaounde have stroke (Njamnshi et al., 2006).
- Recurrence risk without preventive treatment: 60-90% (Powars et al., 1978)

**→ Early detection and treatment are crucial**

# Silent cerebral infarcts (SCI)

- Criteria:

- 1) abnormal MRI
- 2) normal neurological examination
- 3) no history of neurological symptoms

- Difficult to detect clinically

- Frequent (22% before 14 years old) (Miller et al., 2001)

- Associated with increased risk for new SCI and stroke

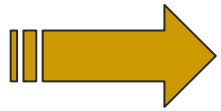
**→ Early detection and treatment is crucial**

# Cognitive consequences of silent infarcts

- IQs of SCD children (Armstrong et al., 1996):
  - Normal MRI: 90
  - SCI: 80
  - Stroke: 70
- Most impaired:
  - Executive functions, attention and visuo-motor coordination (Berkelhammer et al., 2007), difficulties at school
- White et al. (2006):  
neuropsychological battery → 75% accuracy to detect silent infarcts among SCD children

# [ The Project ]

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Contribution of neuropsychological testing to the detection and prevention of stroke in SCD Cameroonian children

# [ Method (1) ]

- Training of 4 Cameroonian psychologists
- Pilot study with 20 subjects
  - ➔ Selection and adaptation of 12 neuropsychological tests

<b>Cognitive Tests</b>	
<b>CVLT-C</b>	<b>Bell Cancellation Task</b>
<b>Purdue Pegboard Test</b>	<b>Continuous Performance Task</b>
<b>Color Trails 1</b>	<b>Digit Span Forward</b>
<b>Color Trails 2</b>	<b>Digit Span Backward</b>
<b>Hand Movements</b>	<b>Letter-Number Sequencing</b>
<b>Verbal Fluency Test</b>	<b>Coding</b>

# [ Method (2) -Norms ]

- Norms: 125 healthy children (71 girls) aged 6 to 20 ( $M=11.4$ ,  $SD=4.2$ ) divided in 7 age-groups (Table 1)

	6-7	8-9	10-11	12-13	14-16	17-20
<b>Boys</b>	8	7	9	8	12	10
<b>Girls</b>	12	13	11	14	10	11
<b>Total</b>	20	20	20	22	22	21

Table 1. Age-groups and gender distribution

- Cut-off scores for each test and age-group

# [ Method(3)-SCD ]

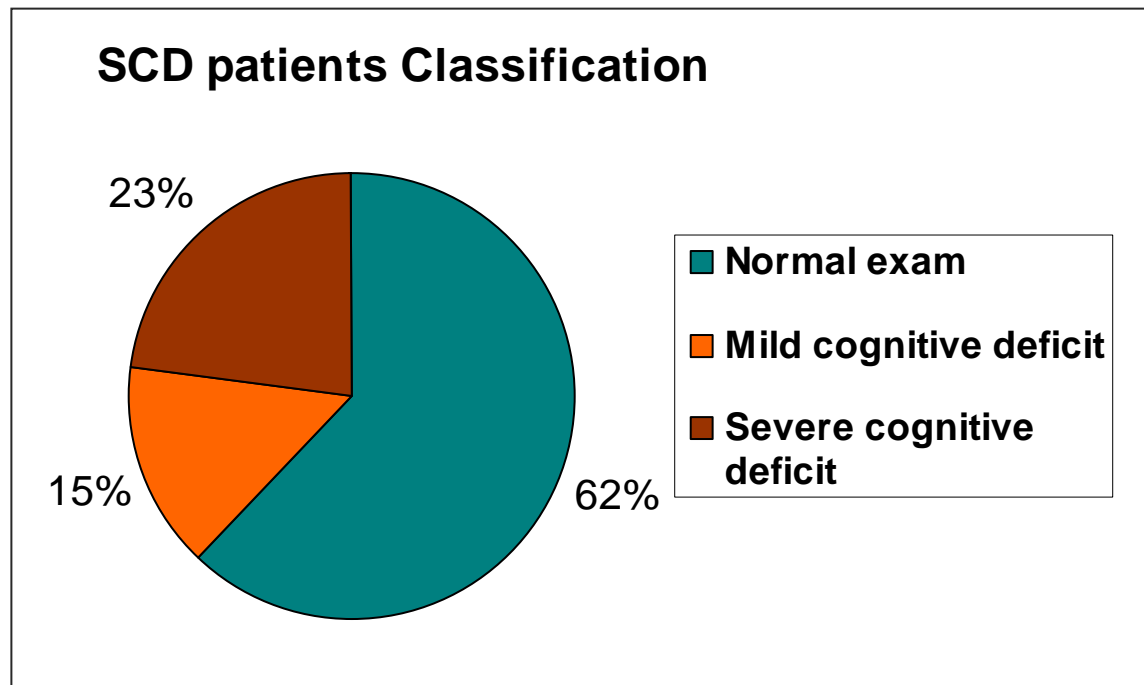
- Classification method:

Classification	Number of test failed (max=12)
Normal exam	0-2
Mild cognitive deficit	3-4
Severe cognitive deficit	>4

- 100 SCD children aged 6 to 24 ( $M=13.4$ ,  $SD=4,9$ )
- Blood test, genetic consultation, neurological exam, Transcranial Doppler and EEG

# [ Results (1) ]

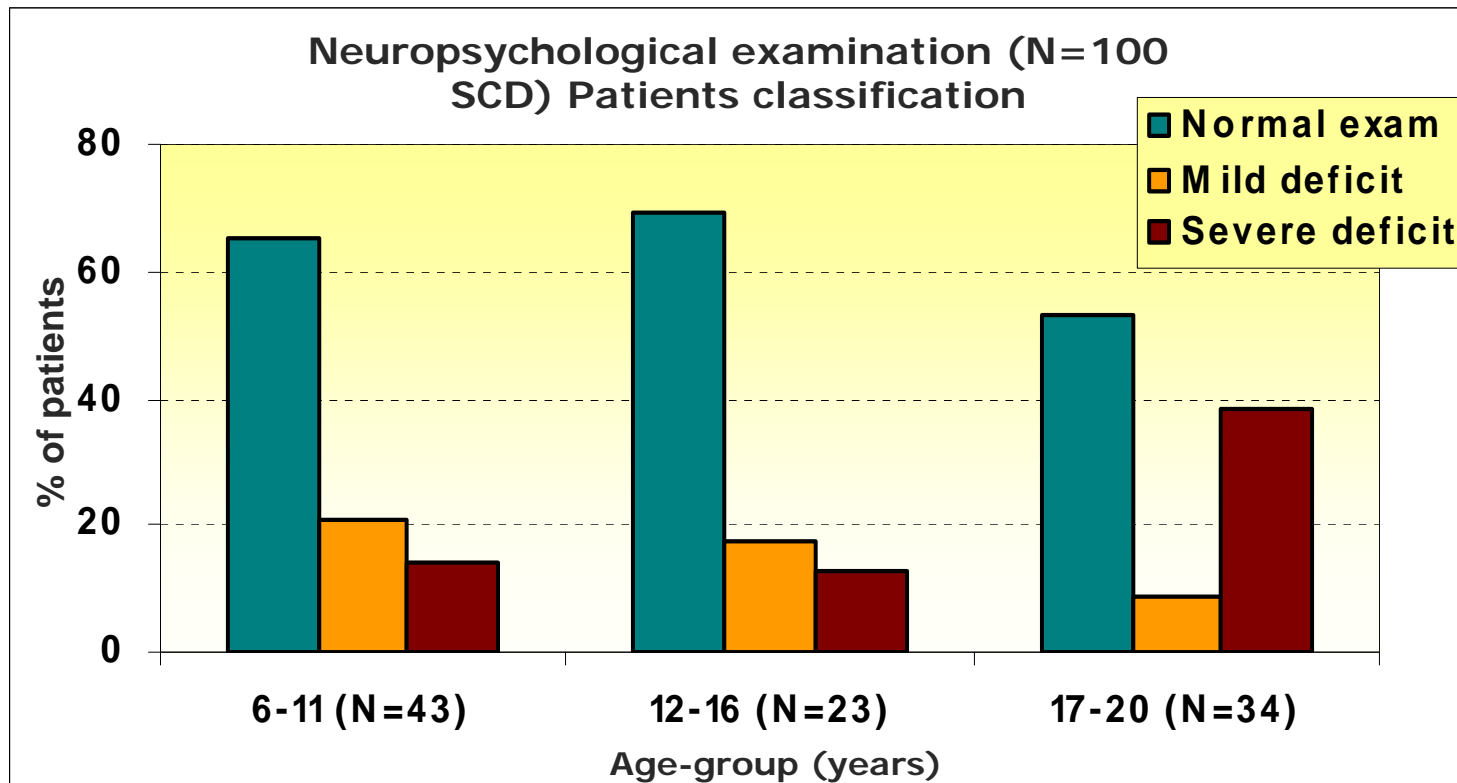
- The overall percentage of SCD children showing mild and severe deficits is high (38%)



# Results (2)

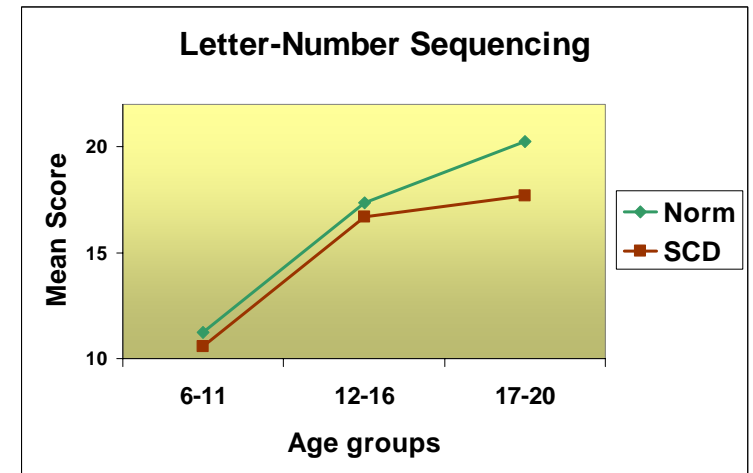
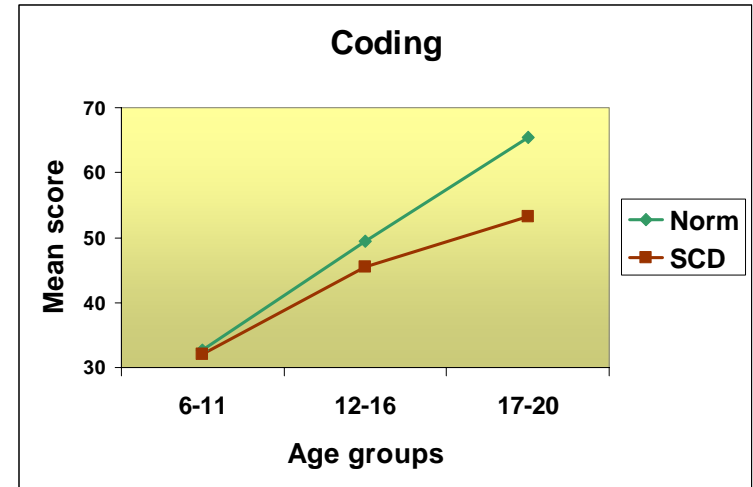
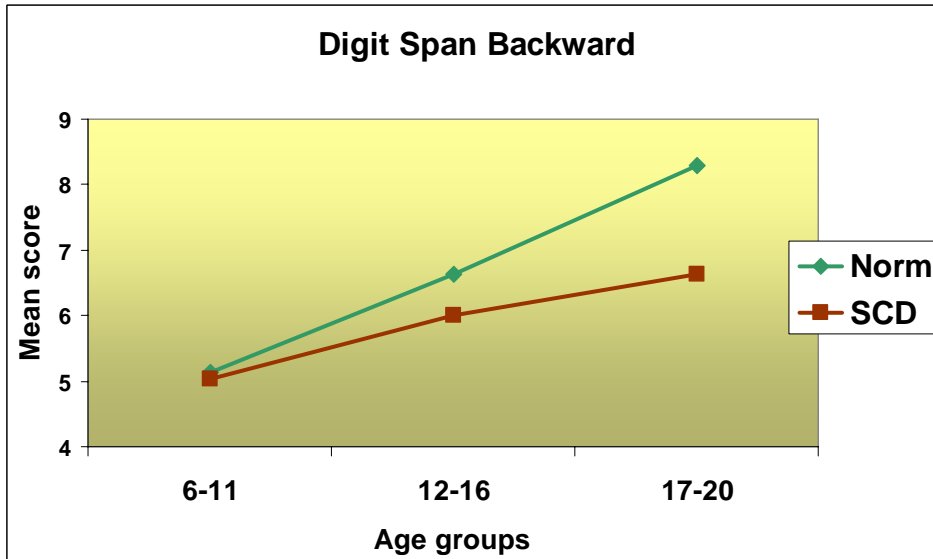
- Evolution of performance with age

➔ SCD children **aged 17 to 20** showed **greater cognitive deficits** than the other groups



# [ Results (3) ]

3. Attention and working memory are most impaired and prototypical of the age related trends



# Conclusions/Perspectives

1. First neuropsychological norms in Cameroon are available and currently used by local psychologists
2. Cognitive function is significantly impaired in Cameroonian SCD children
  - Sickle cell disease seems to have greater negative impact on cognitive function in Cameroonian patients than in western countries
3. Cognitive deficits increase with age
4. Tests assessing attention and working memory are most sensitive to SCD
5. **Future**
  - Follow-up of this cohort over two years to assess the reliability of the battery
  - Enlarge the study sample to younger SCD children (3-5 years old)
  - Enlarge the study population to SCD children followed at the Geneva University Hospital

[ Thank you! ]

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