Early detection and management of extracranial arteriopathy reduces the incidence of silent cerebral infarcts in sickle cell anemia: a long-term prospective cohort study

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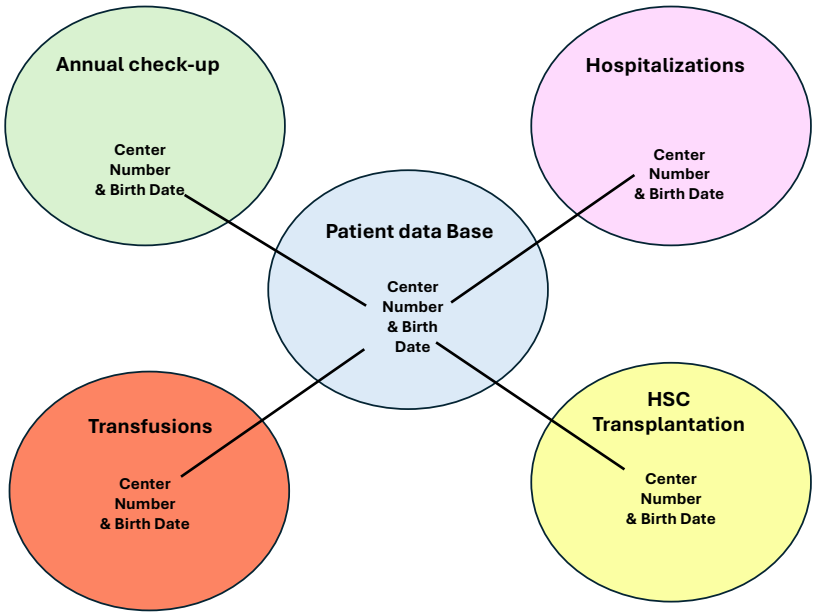
SUPPLEMENTAL FILES

Supplemental Figure 1 .Database organization at the CHIC center
Created in 1992, Logiciel Lotus Approach IBM software, CNIL, N° 2069568
Prospectively updated by the center's physicians
Five databases linked by the center's unique identification number (created on first visit to the center) and by date of birth.



1. **Patient (permanent data)**
   - Center number
   - Birth date
   - Parents origin
   - Date of first visit
   - Diagnosis (genotype)
   - G6PD activity
   - Alpha genes
   - Complete Erythroid phenotype
   - HLA typing
   - Baseline biological parameters recorded in the 2nd year of life before any SCD modifying therapy, 3 months away from transfusion, 1 month from crisis
   - Date of first event: dactylitis, acute splenic sequestration, splenectomy, lithiasis, cholecystectomy, acute chest syndrome, osteonecrosis, transfusion, hospitalization, abnormal TCD, ischemic lesion on MRI, stenosis on MRA, death, transplantation
   - Treatment initiation and discontinuation (hydroxyurea, chronic transfusion, HSCT)

2. **Annual health check**
   a. Clnical data: Height, weight, blood pressure, SaO2, spleen, puberty status
   b. Ongoing treatments
   c. Laboratory data
   d. Imaging data TCD values in each artery, MRI/MRA/neckMRA, cardiac echo, abdominal echo, lung test after age 6, otorinolaryngology, stomatology, ophthalmology examination

3. **Hospitalizations**
   a. Admission and discharge dates
   b. Ongoing treatment on admission (HU?, CT?; HSCT?)
   c. Diagnosis of complication
   d. Transfusion during hospitalization?

4. **Transfusions & Exchanges**
   a. Date
   b. Weight
   c. Hb, HbF, HbS before transfusion
   d. Hb, HbS post transfusion/exchange if necessary
   e. Volume of transfusion

5. **Hematopoietic stem cell transplantation**

a. Date of HSCT
b. Ovary or testis cryopreservation?
c. Allo or gene therapy?
d. Conditioning regimen?
e. HLA-identical? haplo-identical?
f. aGvHD?
g. cGvHD ?
h. Annual Donor chimerism%
i. Annual health check-up

Supplemental Table 1. Silent Cerebral Infarct (SCI) in the overall cohort (n=63)

No	age at first MRI	age at SCI detection	Arteriopathy type	Treatment at SCI detection	Treatment post arteriop/SCI detection	Age at last MRI	Treatment at last MRI
Patients with SCI at first MRI							
1	1.82	1.82	intracranial	No	CT	10.13	CT
2	3.50	3.50	intracranial	No	CT	13.22	HU
3	3.87	3.87	intracranial	No	CT	10.66	alloSCT
4	3.89	3.89	intracranial	No	CT	14.04	CT
5	4.07	4.07	intracranial	No	CT	4.07	CT
6	4.25	4.25	intracranial	No	CT	18.34	HU
7	4.39	4.39	intracranial	No	CT	13.84	alloSCT
8	4.44	4.44	intracranial	No	CT	16.82	CT
9	4.62	4.62	intracranial	No	CT	9.36	alloSCT
10	4.82	4.82	intracranial	No	CT	12.36	alloSCT
11	4.98	4.98	intracranial	No	CT	17.92	HU
12	5.88	5.88	intracranial	No	CT	15.03	HU
13	6.21	6.21	intracranial	No	CT	7.36	alloSCT
14	6.44	6.44	intracranial	No	CT	11.66	HU
15	7.06	7.06	intracranial	No	CT	7.06	CT
16	8.67	8.67	intracranial	No	CT	16.49	alloSCT
17	10.93	10.93	intracranial	No	CT	14.19	CT
18	5.63	5.63	intracranial	HU	CT	7.84	CT
19	6.32	6.32	intracranial	HU	CT	19.59	HU
20	7.37	7.37	intracranial	HU	CT	18.64	CT
21	12.19	12.19	Isolated eICA	No	CT	17.98	CT
22	12.75	12.75	Isolated eICA	No	CT	18.65	CT
23	2.85	2.85	Isolated eICA	No	CT	5.41	CT
24	4.99	4.99	Isolated eICA	No	CT	4.99	CT
25	5.45	5.45	Isolated eICA	No	HU	5.45	HU
26	5.92	5.92	Isolated eICA	No	CT	12.81	HU
27	6.31	6.31	Isolated eICA	No	HU	9.00	HU
28	7.17	7.17	Isolated eICA	No	CT	11.69	CT
29	7.41	7.41	Isolated eICA	No	CT	17.67	HU
30	7.88	7.88	Isolated eICA	No	CT	12.85	CT
31	6.16	6.16	Isolated eICA	HU	CT	12.37	HU
32	3.94	3.94	Isolated eICA	CT	CT	17.56	alloSCT
33	3.28	3.28	absent	No	CT	14.62	alloSCT
34	3.89	3.89	absent	No	CT	15.10	alloSCT
35	4.36	4.36	absent	No	CT	14.75	CT
36	4.42	4.42	absent	No	CT	16.45	alloSCT
37	5.08	5.08	absent	No	CT	14.15	No
38	5.10	5.10	absent	No	HU	8.28	HU
39	5.41	5.41	absent	No	No	7.75	No
40	5.84	5.84	absent	No	CT	9.85	alloSCT
41	9.43	9.43	absent	No	CT	18.31	alloSCT
42	10.03	10.03	absent	No	CT	18.42	HU
43	10.35	10.35	absent	No	CT	17.60	HU
44	13.54	13.54	absent	No	CT	17.73	CT
45	5.64	5.64	absent	HU	CT	16.79	HU
46	10.34	10.34	absent	HU	CT	14.81	HU
47	12.15	12.15	absent	HU	CT	18.22	CT
48	7.14	7.14	absent	CT	CT	16.06	CT
New patients having developed SCI after first MRI, during MRI follow-up							
49	4.34	7.64	intracranial	HU	CT	14.24	HU
50	6.65	9.49	intracranial	HU	CT	12.18	HU
51	5.23	6.61	Isolated eICA	No	CT	10.12	alloSCT
52	3.88	6.94	Isolated eICA	HU	CT	6.94	HU
53	5.58	7.10	Isolated eICA	HU	HU	7.10	HU
54	5.40	9.67	Isolated eICA	HU	HU	9.67	HU
55	4.96	6.45	absent	No	CT	11.50	alloSCT
56	7.40	8.84	absent	No	CT	18.37	CT
57	7.26	9.14	absent	No	No	17.95	No
58	2.78	5.05	absent	HU	CT	6.16	HU
59	5.79	8.15	absent	HU	HU	10.30	HU
60	4.07	10.45	absent	HU	HU	10.45	HU
61	6.87	12.29	absent	HU	HU	17.78	HU
62	10.78	13.10	absent	HU	HU	16.72	HU
63	8.79	17.96	absent	HU	HU	17.96	HU

Footnote: CT: chronic-transfusion, HU:hydroxyurea, No: disease modifying therapy