Follow-up and annual evaluation of children with Sickle Cell Disease General practionner (GP), pediatrician (P) or « specialised» paediatrician (SP)	
At diagnosis	Refer the patient to a specialised paediatrician or team with interdisciplinary structure and comprehensive care
To be performed 8 – 15 days after hospitalization for fever, acute chest syndrome or acute anaemia	Blood counts (GP, P, SP)
Routine evaluation	Every 2 to 3 months if SS or SB° Every 2 to 6 months if SB+ or SC ■ Clinical evaluation (GP, P, SP) ■ Blood counts (GP, P, SP) ■ Hygiene and education (GP, P, SP) ■ Prophylaxis, vaccination, pain treatment at home …(GP, P, SP) ■ Need of disease modifying therapy? (SP) ■ Consider the need to change current chronic treatment (anti-infectious prophylaxis, chelation, Hydroxyurea, chronic transfusion,…) (SP)

Clinical evaluation and EXAMS TO BE PERFORMED ANNUALLY at steady-sate (in the context of a "comprehensive care program")						
	0 – 1 Year	**2 years**	**3 - 5 years**	**6 - 9 years**	**10 – 15 years**	**16 -18 years**
Verify that medical chart contains • Initial Hb electrophoresis (diagnosis) • Basal Hb level and MCV. Basal HbF • G6PD status • Blood group with extended phenotype (at least for Rh, Kell, C and E antigens) • Updated vaccinations						
Physical examination • Growth and development • BMI • Blood pressure • Liver and spleen size at steady-state • Snoaring/ sleep apnoea*** • Etc…						
Transcutaneous O_2 saturation						
Biological tests*						
Indication of disease modifying therapy? Adjustment of HU treatment if indicated (up to MTD if clinically justified) Indication of chronic transfusion/ exchange?						
Adherence (treatments, appointments)						
Parents education (major complication, transmission, prevention, precipitating factors)						
Preparation of surgery if needed						
TCD						
Cerebral MRA if TCD not assessable (no acoustic window) in SS or SB° patients						
Hepatic US						
Hip X-Ray/MRI****						
Electrocardiography Heart echography with Tricuspid Regurgitnt jet Velocity (TRV) If TRV > 2.5 m/sec → right heart catheterisation						

Lung function evaluation if • Asthma • Recurrent ACS • Poor general condition or fatigue • Transcutaneous O2 saturation abnormal						
School success						
Lifestyle and physical and social activities counselling						
Patient education (symptoms, prevention, transmission, precipitating factors) In adolescents: sexuality and contraception						
Ophthalmologic evaluation				**		
Preparation of transition to adult care program						

* Complete blood count, liver profile, electrolytes, BUN, creatinine, microalbuminuria, ferritin if transfused, calcium metabolism including vitamin D and PTH, Parvovirus B19 serology until positive.

** Since the age of 6 y.o. if Hb SC disease

Snoaring/sleep apnoea***: detrimental in SCD. Consider adenoid/ tonsillectomy with appropriate transfusion prior surgery if tonsillectomy

**** MRI more sensitive (to be performed in case of pain and/or limping)

Indications and Requirements for Specific Therapies

Hydroxyurea

Indications

- Established
  - Recurrent episodes of severe acute pain ≥ 2-3 per year requiring hospitalization?
  - ≥2 episodes of acute chest syndrome
- Postulated
  - Correction of severe anemia
  - Primary Stroke prevention
  - Prevention of organ dysfunction

Monitoring

- Full blood count and Hb F level each 2 weeks after initiation and after each dose increase; when stable every 8 weeks
- Monitor spleen size, particularly if splenomegaly is present or there is an history of splenic sequestration
- Propose storage of frozen sperm (?)

Red cell transfusion

- Indications
  - Prevention of cerebrovascular events (primary and secondary prevention)
  - Response failure to hydroxyurea for acute chest syndrome or recurrent painful crisis
  - Recurrent splenic sequestration (children < 5 y.o.). Hydroxyurea may be also an option. Splenectomy < 5 y.o. to consider also (asplenia already present in very young children) with appropriate prophylaxis
  - Prevention of organ dysfunction if response failure to HU

- Prevention of alloimmunization and iron overload
    - Phenotypic matching for full Rh, Kell antigens, and more extensively if alloimmunization is known or suspected
    - Chronic exchange transfusions to be preferred to top-up transfusions
    - Iron chelation should be considered for patients who have received at least 20 top-up transfusion episodes or with a serum ferritin level consistently >1,000 µg/L