Early splenectomy in a large cohort of children with sickle cell anemia: risks and consequences

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Supplemental data

Cases of splenectomy < 2 years

- First case was a child who had a 2nd attack of acute splenic sequestration while on a chronic transfusion program in addition to an unfavourable socio-economic situation that would not allow the family to urgently seek medical care in case of a 3rd recurrence.
- 2d case was a child on a chronic transfusion program who presented with delayed haemolytic transfusion reaction with acute splenomegaly.
- 3rd case was a child who presented a 4th episode of acute spleen sequestration with threatening anaemia (Hb 2.8 g/dl)

Supplemental Table 1: Indications for hematopoietic stem cell transplant in the cohort of splenectomized children

Patients	Age at HSCT (years)	Indication
1	11,96	severe symptomatic disease with multiple VOEs despite hydroxyurea necessitating chronic transfusion therapy
2	9,88	severe symptomatic disease despite hydroxyurea + alloimmunization (post splenectomy)
3	11,78	Symptomatic disease + allo immunisation (pre splenectomy)
4	13,18	Delayed Haemolytic Transfusion Reaction
5	3,98	Cerebral vasculopathy (abnormal TCD)
6	12,35	severe symptomatic disease with multiple VOEs despite hydroxyurea
7	7,44	Renal insufficiency related to malformative uropathy
8	7,12	Cerebral vasculopathy (abnormal TCD)
9	3,08	acute splenic sequestration

VOE: vaso occlusive event; TCD : Trans cranial Doppler;

Cases of splenectomy following one single episode of acute splenic sequestration (ASS):

- Case 1: severe ASS with acute threatening anaemia (Hb= 3,3 g/dl.)
- Case 2: severe ASS with acute threatening anaemia (Hb=1,5 g/dl.)
- Case 3: severe ASS with acute threatening anaemia (Hb= 4.7 g/dl.) and rare blood group phenotype (S-s-) and allo antibodies
- Case 4: severe ASS with acute threatening anaemia (Hb= 3,4 g/dl.)