P1471 CURRENT INDICATIONS AND RISKS OF SPLENECTOMY IN A LARGE MODERN COHORT OF CHILDREN WITH SCA

Topic: 26. Sickle cell disease

Aimen Mechraoui1, Ghislaine Ithier1, Justine Pages1, Zinedine Haouari1, Liza Ali3, Arnaud Bonnard3, Malika Benkerrou1, 4, Florence Missud1, Berengère Koehl1, 5, 6, Laurent Holvoet1, Enora Le Roux1, 4, Valentine Brousse1, 5, 6

1 Site de référence labellisé sur la drépanocytose, filière MCGRE, Service d’Hématologie-ImmunoLogie, Hôpital Universitaire Robert Debré, Paris, France; 2 Unité d’épidémiologie clinique, Inserm, CIC 1426, F-75019, Hôpital Universitaire Robert Debré, Paris, France; 3 Service de Chirurgie, Hôpital Universitaire Robert Debré, Paris, France; 4 Inserm, UMR-1123 ECEVE, Université de Paris, Paris, France; 5 Biologie Intégrée du Globule Rouge, Unité Mixte de Recherche S1134, INSERM, Université de Paris, Paris, France; 6 Laboratoire de Référence GR-Ex, Université de Paris, Paris, France

Background:

In children with sickle cell anemia (SCA), the spleen is altered early in life and may be the site of serious complications such as acute splenic sequestration (ASS) or hypersplenism. Surgical splenectomy may be necessary in those who experience such complications, but both the consequent infectious and thrombo-embolic risks, and the age at which splenectomy may be safely performed, remain unclear.

Aims:

The objective of our study was to determine the post-splenectomy incidence of invasive bacterial infections and thrombo-embolic complications in a large cohort of children with SCA. The secondary objective was to analyze the effect of age at splenectomy on these risks.

Methods:

Retrospective review of children with SCA (SS or SB0-thalassemia) splenectomized during the 2000-2018 period at Robert Debré University Hospital (Paris, France) was undertaken. The study received ethical approval. The main clinical characteristics of patients, indications for splenectomy and outcomes were analyzed.

Results:

A total of 188 children were included, with 164 (87.2%) SS children. Median (Q1-Q3) age at splenectomy was 4.1 yrs (2.5-7.3). Median duration of post splenectomy follow-up (FU) was 5.9 yrs (2.7-9.2) yielding 1192.6 patients-years (PY) of observation. One death (0.5%) occurred during FU, unrelated to splenectomy. Indications for splenectomy were ASS in 101 (53.7%) cases, hypersplenism in 75 (39.9%) and diverse reasons in the remaining children (splenomegaly n=5; delayed hemolytic post transfusion reaction n=5, other n=2). Pneumococcal immunization coverage was good with 166 (95.9%) eligible children immunized with polysaccharidic pneumococcal vaccination at the time of splenectomy and 7 (4.0%) after the procedure. Regarding the conjugated pneumococcal vaccination, 158 (91.9%) of eligible patients had received at least one injection at the time of splenectomy. All patients received penicillin prophylaxis. Overall incidence of invasive bacterial infection was 0.005/PY (n=6 in 5 patients, no pneumococcal infections). Overall incidence of thrombo-embolic events was 0.003/PY (n=4 in 4 patients).

In this cohort, 123 (65.4%) and 65 (34.6%) children were splenectomized > or < 3 years of age, respectively. Overall incidence of invasive bacterial infection and thrombo-embolic events was not different according to the age at splenectomy (0.005/PY in both groups) and 0.004/PY versus 0.003/PY (p= 0.7), respectively. Likewise, there was no difference between these groups in the incidence of acute chest syndrome or vaso-occlusive events. Conversely, a
significant increase in the overall incidence of cerebral vasculopathy (including abnormal TCD, stroke and cerebral stenosis) was found in children splenectomized before 3 (0.037/PY) versus after 3 (0.011/ PY), p<0.01).

Patients who presented with ASS (n=101) were splenectomized at a median age of 3.3 yrs (2.4-5.4), following a median number of 3 (2-4) episodes of ASS and therefore contributed to a large proportion of patients splenectomized under 3 (n=44/65, 67.7 %).

**Summary/Conclusion:** To date this is the largest pediatric cohort study of splenectomized patients with SCA. We show that surgical splenectomy does not result in a significant increased risk of complications, notably invasive bacterial infections or thrombo-embolic complication. Splenectomy should not be delayed in patients once there is an indication for surgical removal, and the children have received recommended vaccines and prophylactic penicillin therapy. The relationship between ASS, splenectomy and cerebral vasculopathy needs to be further assessed.