LETTER TO THE EDITOR

Morbidity and mortality of sickle cell disease patients is unaffected by splenectomy: evidence from three decades of follow-up in a high-income setting

Valeria Maria Pinto,[1*] Barbara Gianesin,[2*] Frédéric B. Piel,[3] Filomena Longo,[4] Paolo Rigano,[5] Alessandra Quota,[6] Vincenzo Spadola,[7] Giovanna Graziadei,[8] Filippo Mazzi,[9] Maria Domenica Cappellini,[8] Aurelio Maggio,[5] Antonio Piga,[10] Lucia De Franceschi[9#] and Gian Luca Forni[1#]

[1]Center for Microcythemia, Congenital Anemia and Iron Dysmetabolism, Galliera Hospital, Genoa, Italy; [2]ForAnemia Foundation, Genoa, Italy; [3]Department of Epidemiology and Biostatistics, School of Public Health, Imperial College London, London, UK; [4]Reference Center for Hemoglobinopathies, AOU San Luigi Gonzaga Hospital, Orbassano, Italy; [5]Campus of Hematology Franco and Piera Cutino, AOOR Villa Sofia-V. Cervello, Palermo, Italy; [6]Thalassemia Unit, P.O. Vittorio Emanuele III, Gela, Caltanissetta, Italy; [7]Thalassemia Center, P.O. Giovanni Paolo II, Ragusa, Italy; [8]Department of Medicine and Medical Specialities, IRCCS Ca' Granda Foundation, Maggiore Policlinico Hospital, Milan, Italy; [9]Department of Medicine, University of Verona & AOUI Verona, Policlinico GB Rossi, Verona, Italy and [10]Department of Clinical and Biological Sciences, University of Turin, Turin, Italy.

VMP and BG contributed equally as co-first authors.
#LDF and GLF contributed equally as co-senior authors.

Correspondence: GIAN LUCA FORNI - gianluca.forni@galliera.it

https://doi.org/10.3324/haematol.2022.280815

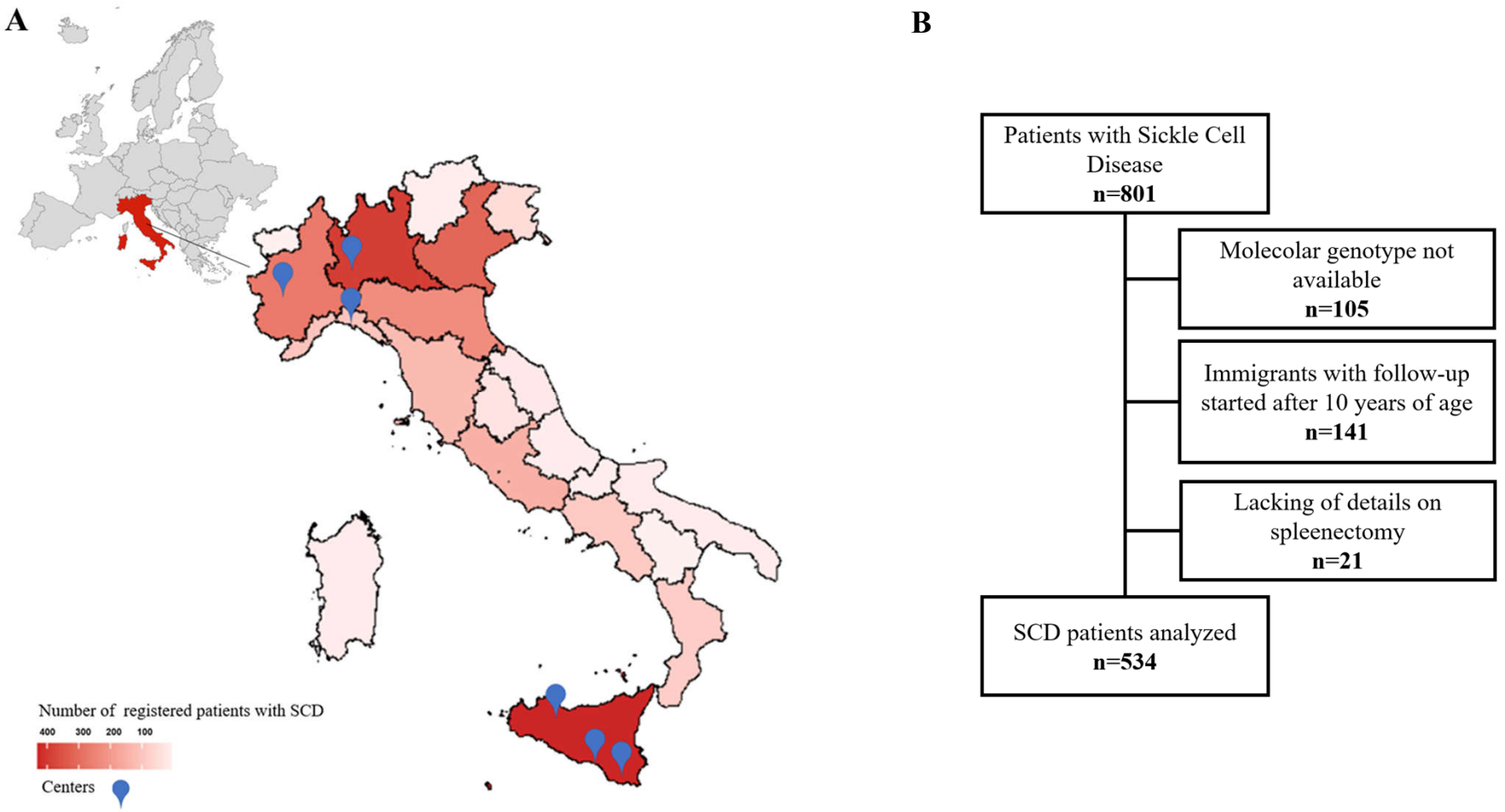

Figure S1. A. Distribution of sickle cell disease (SCD) in Italy and comprehensive centers for hemoglobinopathies with long-term followed-up patients with SCD patients (n=2,300). **B.** Flow-chart of study population (SCD: Sickle Cell Disease).

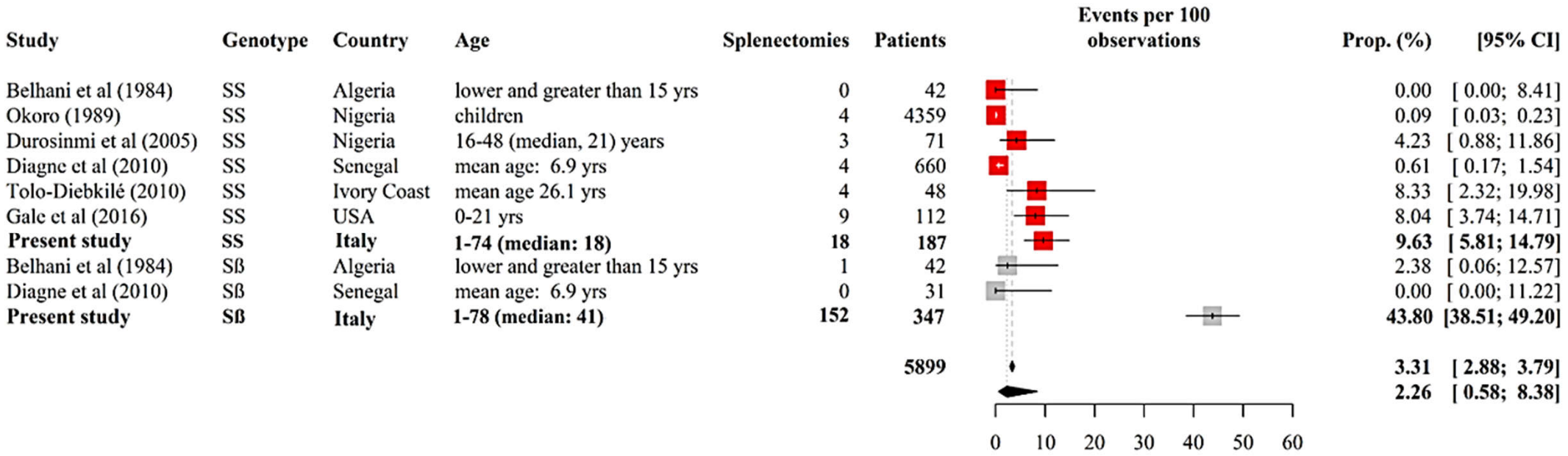

Figure S2. Forest plot of observational studies reporting cases of surgical splenectomy in SCD for genotype SS (red) and Sβ, considered as Sβ ° plus Sβ+ (gray). The studies reported were included in the systematic review of Ladu et al. (2021).

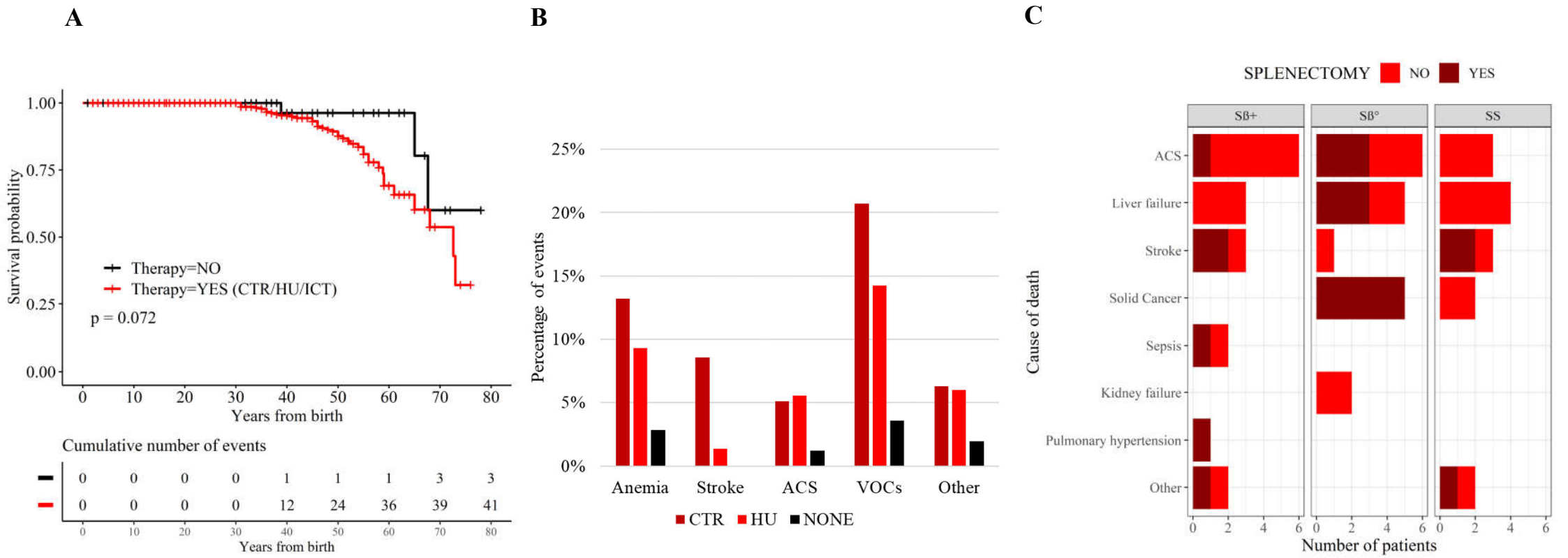

Figure S3. A. Survival probability for patients without therapy (NO) or treated with CTR, HU or ICT therapy. **B.** Percentage of sickle cell related events occurred in SCD patients by treatment type (HU/CTR/NONE). (CTR: chronic transfusion regimen; HU: hydroxyurea; ICT: iron chelation therapy). **C.** Causes of death in splenectomized (YES) or not-splenectomized (NO) patients with sickle cell disease according to genotypes