



The 65th ASH Annual Meeting Abstracts

POSTER ABSTRACTS

113. SICKLE CELL DISEASE, SICKLE CELL TRAIT AND OTHER HEMOGLOBINOPATHIES, EXCLUDING THALASSEMIAS: BASIC AND TRANSLATIONAL

Antinuclear and Specific Autoantibodies Predict Autoimmune Disease in Sickle Cell Patients: A Longitudinal Cohort Study

Frederic Truffinet¹, Caroline Compain², Maria Elena Manea², Anoosha Habibi³, Elena Fois², Etienne Audureau^{4,5}, Sophie Hue⁶, Marc Michel⁷, Pablo Bartolucci⁸

¹CHU Henri Mondor. UPEC. AP-HP, Paris 12, France

²CHU Henri Mondor. UPEC. AP-HP, Creteil, France

³sickle cell referral center, APHP, Hôpital Henri Mondor, Creteil, France, Joinville le pontCréteil, France

⁴Henri Mondor APHP, Creteil, France

⁵Unité de Recherche Clinique, Henri-Mondor University Hospital- UPEC, AP-HP, Creteil, France

⁶CHU Henri Mondor. UPEC. AP-HP, creteil, France

⁷Internal medicine department, Henri Mondor Hospital, APHP, Créteil, France

⁸CHU Henri Mondor. UPEC. AP-HP, Créteil, France

Introduction: Sickle cell disease (SCD), the most frequent genetic disorder, can be associated with systemic autoimmune diseases (ADs). High levels of antinuclear antibodies (ANA) are commonly described in SCD, as alloimmunization after blood transfusion. However, the specificity and the prognostic value of ANA have not been fully determined.

We aimed to evaluate the association of ANA with AD development during long-term follow-up in SCD patients. Our secondary goal was to assess mortality and serious SCD-related events in this population.

Patients and methods: We conducted a single-center longitudinal cohort study on 66 adult SCD patients with ANA titers $\geq 1/320$ at baseline followed over a 13 year-period. Patients known to have an AD (n=9) were excluded from the risk analysis.

Results: During follow-up, 13 out of 57 patients (23%) developed AD. Patients with ANA titers $\geq 1/1280$ had a 3-fold increased risk of developing AD (RR 3.4[1.4;8.6]) and a 9-fold increased risk in the presence of specific autoantibodies (anti-DNA, anti-ENA, RF or ACPA) (RR 9.0[3.7;23]) with a positive (PPV) and negative predictive value (NPV) of 90%. When these two immunological features were associated, specificity and PPV reached 100%, with a NPV of 84%. There was no significant difference in the occurrence of serious SCD-related events or mortality rate according to AD.

Conclusions: Sickle cell patients with high level of antinuclear antibodies and specific autoantibodies are at high risk of developing AD. The occurrence of AD does not seem to have a significant impact on the course and severity of SCD.

Disclosures Michel: UCB: Honoraria; Sobi: Consultancy; Novartis: Consultancy; Alexion: Consultancy; argenx: Honoraria; Sanofi: Consultancy.

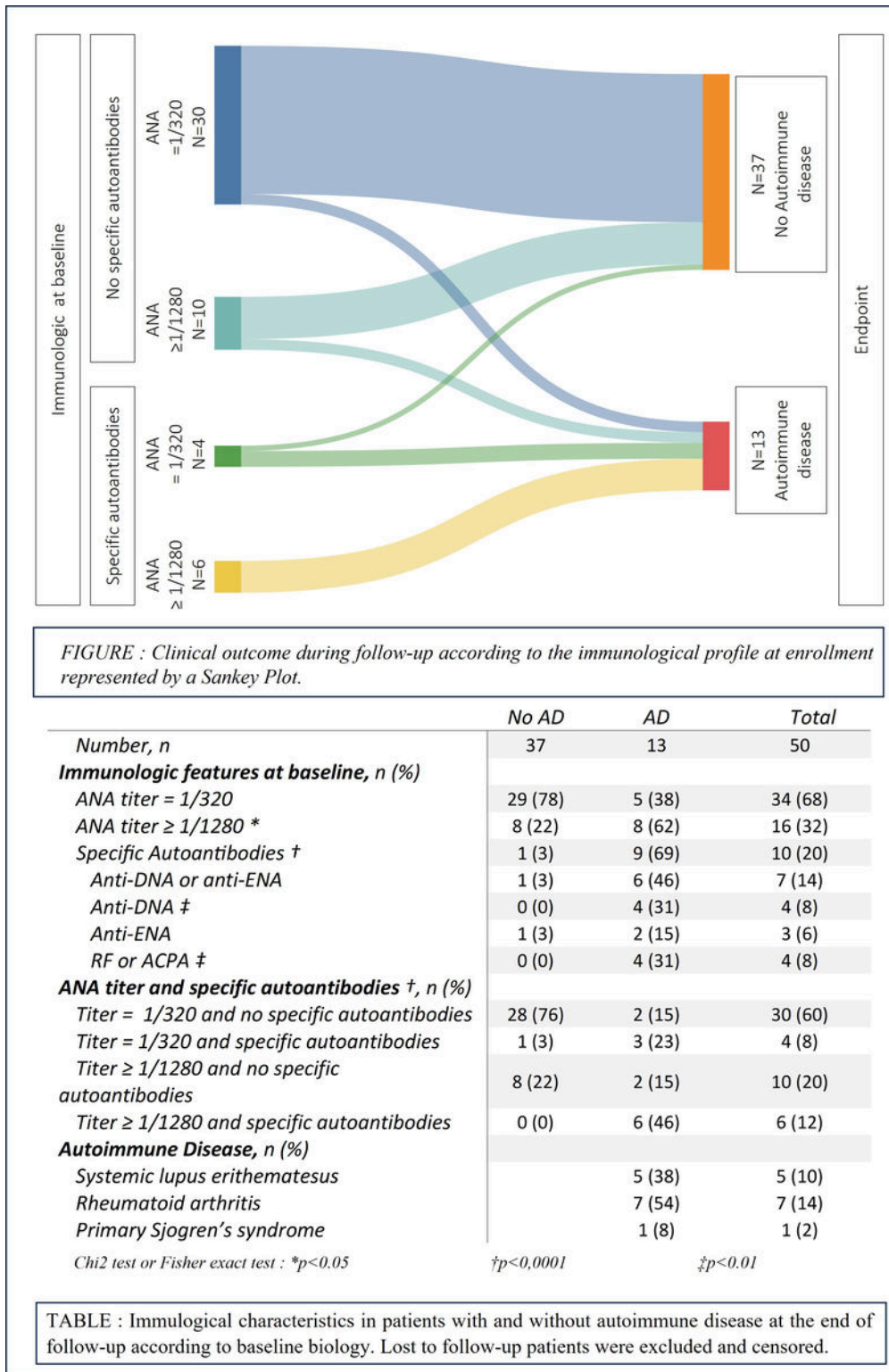


Figure 1

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