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ABSTRACT N. CO33

MALATTIE EMORRAGICHE CONGENITE E ACQUISITE

# ACQUIRED HEMOPHILIA A IN LIGURIA, ITALY: WHAT SIX YEARS OF DATA REVEAL.

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Background and Aims: Acquired hemophilia A (AHA) is a rare autoimmune bleeding disorder caused by the development of autoantibodies that target various epitopes on the factor VIII (FVIII) molecule. These autoantibodies inhibit FVIII's coagulation activity and accelerate its clearance. AHA remains a significant challenge for clinicians, as many physicians often poorly recognize it. This hampers its diagnosis and likely leads to an underestimation of both its prevalence and incidence.

The Liguria Region has a total population of 1,565,307 people thus the estimated incidence of new diagnoses of AHA is 2.3 cases per million. Our aim was to analyze the cases of AHA recorded from January 2018 to October 2024 in the Liguria region, focusing on epidemiological, clinical-therapeutic, and organizational aspects.

Methods: We retrospectively analyzed data from patients with acquired hemophilia A admitted at IRCCS Policlinico San Martino (Genoa) between Jan 2018–Aug 2024. Inclusion criteria were FVIII:C <50% and inhibitor >0.6 BU/mL. Diagnosis was based on recent bleeding, prolonged aPTT, and reduced FVIII, confirmed via mixing test and Nijmegen-modified Bethesda assay.

Results: 22 patients (10F, 12M; median age 80) were found. Estimated incidence was 2.3 cases/million. Most diagnoses (21/22) occurred in Liguria's ASL 1–3. Common comorbidities included autoimmune diseases (n=8) and MGUS (n=3); only 2 patients had neoplastic conditions. Median time to diagnosis was 4 days. No correlation was found between bleeding type, FVIII levels, or inhibitor titers. Anti-porcine inhibitors were positive in 3/10 tested. All patients received immunosuppressive therapy (IST, 5 regimens), achieving 100% complete remission; median time to inhibitor clearance was 20 days. Relapse rate was 31%, all managed successfully with second-line IST (median 8 days). A second relapse occurred in 13% (median 114 days). Infectious complications arose in 31%. Hemostatic treatment was required in 91%; 47% of those treated with bypassing agents (BPAs) needed switching to rpFVIII, which proved effective. One patient received rpFVIII as first-line therapy with no need for further intervention (Figure).

Conclusions: We found an average incidence of 2.44 cases per million, but with a very uneven distribution. Therefore, assuming a proportional number of undiagnosed cases in areas where no diagnoses have been reported, we would estimate an incidence of at least 3.6 cases per million. Most importantly, we should consider the possibility of around ten undiagnosed cases, potentially resulting in at least two deaths. This outlines the need for higher awareness and diagnostic capabilities in the eastern areas that is possible to reach with adequate training for medical and nursing staff particularly in emergency departments especially for peripheral hospitals. Such training could allow faster diagnosis, prompt treatment, better outcome, shorter hospital stay and ultimately lower costs.

We noticed the need for a more standardized approach to the IST of AHA that would facilitate the comparison of different IST regimens, enabling the identification of the most effective treatment strategies tailored to the specific needs of each patient.

Finally, susoctocog α, especially at a loading does of 100IU/kg, seems to be the most effective treatment for bleeding control with the added value of a lower thrombotic risk, already increased in elderly patients.

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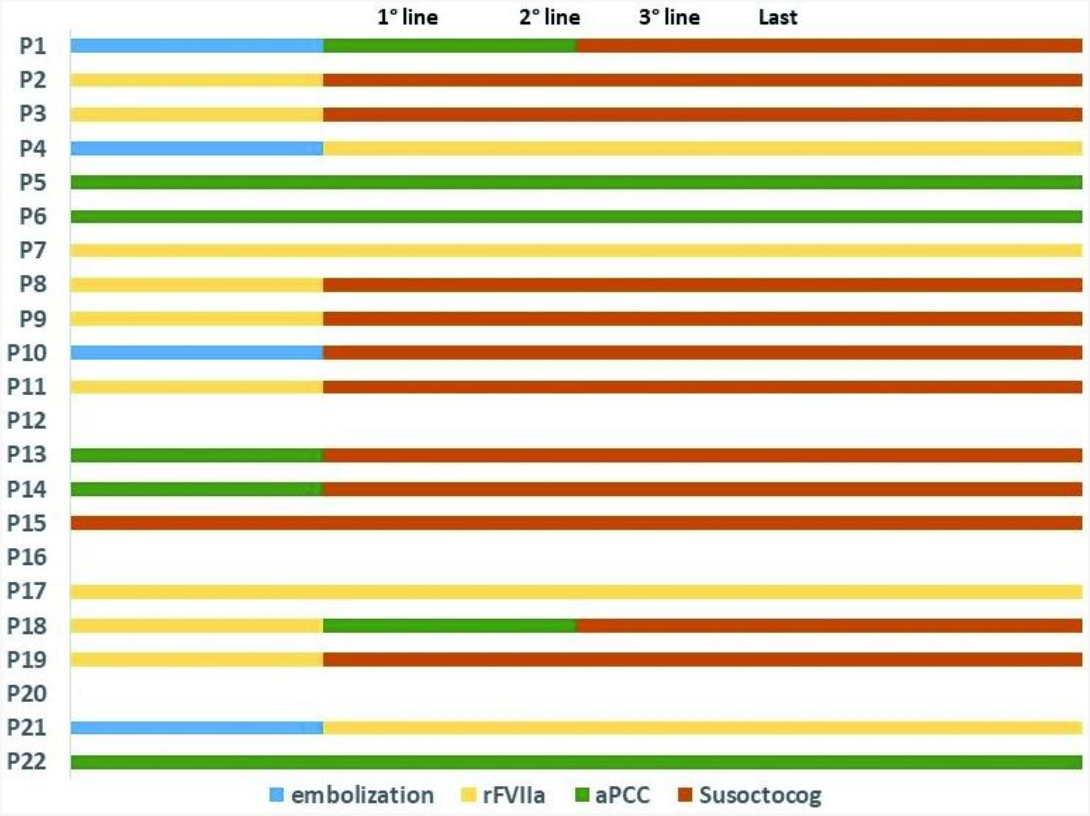
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