

option, especially for patients with cytopenias and no neuropathy. BR is an effective chemotherapy regimen for WM, but myelotoxicity can be an issue in already cytopenic patients. Ibrutinib with or without rituximab can be used in first line or relapsed patients, but it seems to have a worse response in patients with wild type MYD88. Acalabrutinib and Venetoclax are other new options with response in relapsed setting, including in patient's refractory to ibrutinib. Zanubritinib, a new BTK inhibitor for WM treatment, is at least as effective as ibrutinib with perhaps a better toxicity profile.

Marginal Zone B-cell lymphomas (MZL)

The marginal zone B-cell lymphomas (MZLs) comprise extra nodal MZL (EMZL) of mucosa-associated lymphoid tissue (MALT), splenic MZL (SMZL) with or without villous lymphocytes and nodal MZL (NMZL) with or without monocytoid B cells. These are three distinct clinical entities with specific diagnostic criteria, clinical and therapeutic implications.

Regarding to localize H. pylori-positive gastric MZL, the initial treatment should be H. pylori eradication. This treatment can induce lymphoma regression and long-term clinical disease control in the most of 50% of the patients. In patients who do not achieve lymphoma regression following antibiotic therapy, and the rare ones not associated with H. pylori infections, radiotherapy seems to be the best choice (considering localized disease). Patients who require systemic treatment are not very common, but long-term data from the randomized study IELCS-19 showed better response rates and event-free survival when adding rituximab to chlorambucil, but no OS gain compared to chlorambucil alone⁵. SMZL in asymptomatic patients should be observed. There are no randomized trials, but when treatment is required, splenectomy and rituximab monotherapy are considered first-line. For asymptomatic patient diagnosed with NMZL is also recommended only observation. If systemic treatment is indicated, chemoimmunotherapy can be performed.

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ELN recommendations for treating MDS

David Bowen

Whilst peer-reviewed published guidelines have appropriate scrutiny and endorsement to support their validity, the reality is that these often become at least partially outdated soon after publication. In developing the latest iteration of guidelines for the diagnosis and management of MDS, the European LeukemiaNet MDS guideline group, has sought to create a web-based interface that is interactive, portable, and capable of dynamic updating at least annually. The guideline development process involved systematic literature review, expert opinion, and scenario analysis. The faculty was diverse; from 18 European countries. The group included Junior Faculty who fed back on content and the practicality for access from mobile devices. The final product is interactive and iterative, with upfront 'headline' recommendations supported by expanded information pages for readers requiring further detail [<https://mds-europe.org/>]. Examples of updates from our previous ELN guidance are:

Diagnosis: we suggest mutation analysis in all patients where available, to inform prognosis and management. We explain the strengths and the limitations of current knowledge, including discussion of the clonal cytopenias. We also include consideration of germline predisposition syndromes.

Prognosis: we provide calculation tools for IPSS-R and will update this with the forthcoming IPSS-Mol during late 2021/2022.

Low-risk MDS: we describe new data for iron chelation, for early use of Erythropoietic Stimulating Agents and for novel agents such as Luspatercept. Pathways for use of these agents are presented.

High-risk MDS: recommendations are given for the use of hypomethylating agents, chemotherapy, and allogeneic stem cell transplant. An interactive stem cell transplant algorithm including comorbidity is available to guide transplant decisions.

This project was supported by Horizon 2020 funding under the auspices of the MDS-RIGHT programme. The guidelines have recently been endorsed by the European Haematology Association. Discussions are ongoing with international colleagues such that the website may accommodate international variation of recommendations.

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How I treat Polycythemia vera

Barbara Mora, Francesco Passamonti

Polycythemia vera (PV) is a Philadelphia-negative myeloproliferative neoplasms (MPNs),^[1] characterized by high myeloid cells production secondary to mutations in Janus kinase 2 (JAK2) gene.^[2,3] Incidence rate is higher in advanced age.^[4]