## Form record received

International Workshop on Waldenstrom's Macroglobulinemia <pattersonkent@outlook.com>
Mon 7/15/2024 4:02 PM
To:Patterson, Christopher <Christopher_Patterson@DFCI.HARVARD.EDU>

External Email - Use Caution

Record saved to database with ID: 150

Form ID: 1
Form title: Abstract Submission
Form name: Abstract_Submission
Submitted at: 2024-07-15 15:59:08
Submitter IP: 176.132.218.230
User-ID: 0
Username: -
User full name: -
Submitter provider: Unknown
Submitter browser: Mozilla/5.0 (Macintosh; Intel Mac OS X 10_15_7) AppleWebKit/605.1.15 (KHTML, like Gecko) Version/17.3 Safari/605.1.15
Submitter operating system: mac

First Name: EMILIA-LARISA
Last Name: ZIDARU
Email: larisazidaru@yahoo.com
Phone Number (optional): +40768667916
Registration Type: Delegate
Abstract Title: Single-Center Experience, Clinical Spectrum, and Prognosis of Patients with IgM AL Amyloidosis
Select abstract file to attach:
/home/dkwolfpk2016/public_html/waldenstromsworkshop/media/breezingforms/uploads/abstract2024.docx
Conference: IWWM12

# Single-Center Experience, Clinical Spectrum, and Prognosis of Patients with IgM AL Amyloidosis

S. Badelita[1], Sinziana Barbu[1], L. Zidaru[1], A. Jercan [1], M. Draghici [1], M. Popescu , C. Dobrea, D. Coriu [2]
1 Fundeni Clinical Institute, Bucharest, Romania;
2 Carol Davila University of Medicine and Pharmacy, Bucharest, Romania

**Introduction:** IgM-associated light chain amyloidosis (AL) accounts for 5-7% of all systemic AL cases and occurs in 7.5% of patients with Waldenström's Macroglobulinemia (WM)/lymphoplasmacytic lymphoma (LPL). Patients with IgM AL exhibit more soft tissue, lung, and peripheral nerve involvement but less cardiac involvement and generally have worse outcomes compared to non-IgM AL patients due to lower hematologic response rates.

**Aim:** This retrospective, single-center study aims to delineate the characteristics and progression-free survival (PFS) of patients diagnosed with IgM amyloidosis.

**Methods:** Between January 2010 and December 2023, 214 patients were diagnosed with AL Amyloidosis at the Fundeni Clinical Institute in Bucharest, Romania. Among these, 13 patients were diagnosed with IgM AL Amyloidosis, confirmed through Congo Red staining and immunohistochemistry. Organ involvement and treatment response were assessed using consensus criteria with IgM AL prevalence calculated at 6.1%.

**Results:** The median age at diagnosis was 61 years, with a time to diagnosis at 12 months. Kappa light chains predominated in 61% of patients, while 39% had lambda light chains, with median levels of 88.2 mg/L for kappa and 166 mg/L for lambda. FISH examination, performed in 2 cases, revealed del 17p mutation in 1 patient. MYD88 testing was positive in 2 out of 6 patients. Mayo staging showed 38% of patients in stage II, 7.6% in stage I, 7.6% in stage III, with 46% unevaluable.

Renal involvement was most common, seen in 69% of patients, followed by lymph node and peripheral nervous system involvement (38%), cardiac and autonomic nervous system involvement (30%), and splenic involvement (15%). No hepatic or gastrointestinal involvement was noted.

Most patients (69%) were treated with anti-CD20 monoclonal antibody-based therapies (rituximab), with 30% receiving BDR, 15% receiving R-Bendamustine, 15% R-CVP, and 7% R-Ibrutinib. MP and VCD regimens were each administered to 2 patients (15%). Hematologic overall response rate (ORR ≥ VGPR) was achieved in 69%, partial response (PR) in 23%, and 1 patient's response was unevaluable. Despite good hematologic responses, organ responses were seen in only 46%, with 38% renal and 7.6% cardiac responses.

PFS after first-line treatment was 37 months, with a notably higher PFS in patients with exclusive renal involvement (76 months) compared to those with >3 organ involvements (23 months). Death was recorded in 53% of patients

**Conclusions:** Patients with IgM AL amyloidosis in this study exhibited a diverse range of organ involvement, with renal involvement being the most common and having the greatest impact on outcomes. Renal involvement was associated with better progression-free survival (PFS) compared to other organ involvements. Although hematologic responses were common, organ responses were limited, underscoring the poor prognosis for these patients.