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Heart Failure

BORTEZOMIB/DEXAMETHASONE-BASED TREATMENT FOR LIGHT-CHAIN CARDIAC AMYLOIDOSIS RESULTS IN MARKEDLY PROLONGED SURVIVAL COMPARED TO PREVIOUSLY USED REGIMENS

Oral Contributions
South, Room 104
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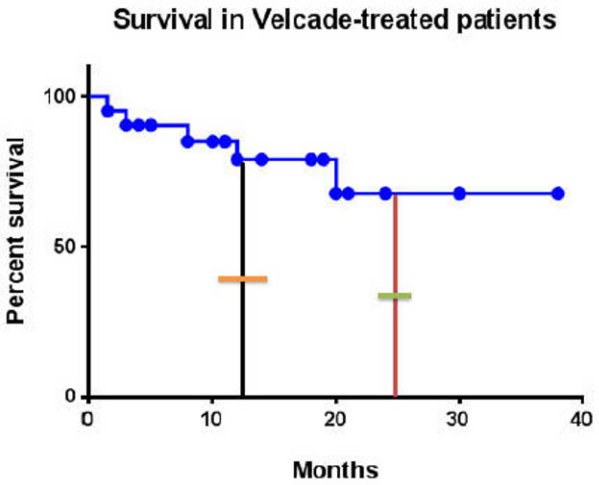
Background: Light chain (AL) amyloidosis is a plasma cell

disease related to myeloma. Progression of AL cardiac amyloidosis (CA) is due to amyloid infiltration and light-chain toxicity. Untreated, CA has a median survival of 6 months, which can be prolonged to 12 month with melphalan. Bortezomib (BORT), a proteasome inhibitor, rapidly reduces circulating free light chains, prolongs disease-free survival in myeloma and has minimal cardiotoxicity. We report our experience with a BORT-dexamethasone regimen in previously untreated patients with AL CA.

Methods: 21 consecutive patients, mean age 62.4+/-9yr (12 men) with CHF due to AL amyloidosis were treated with IV BORT given weekly with 10-40 mg dexamethasone. 8 patients also received cyclophosphamide. 14 had lambda predominant disease and 7 kappa. 12 had disease isolated to the heart. Response to therapy was based on decrease in serum free light chains and survival.

Results: 18/21 showed hematologic response, with normalization of light chains (causing cessation of amyloid deposition) in 12. Clinical improvement occurred in 16 patients and major, reversible side-effects were seen in 5. Actuarial survival was 80% at 1 year and 70% at 2 years and median survival had not been reached.



Vertical lines show 12 and 24 month survival
Horizontal bars show corresponding survival with melphalan, based on published data

Conclusions: BORT/steroid based treatment for AL CA is a highly effective therapy. Administration requires a skilled cardio-oncologic team, but produces marked survival benefit in a previously rapidly progressive disease.