Nine patients were male, 14 patients were female. Their mean age at diagnosis was 71 years (range, 57 to 81 years). None of the patients had iron deficiency or a post-splenectomy state. The median follow-up from the time of diagnosis was 10.8 years for the JAK2-V617F positive group and 9.1 years for the negative group. Arterial events included stroke (n=2; JAK2-V617F negative), transitory ischemic attack (n=1, JAK2-V617F negative) and peripheral arterial thromboses (n=2; JAK2-V617F positive). Deep vein thrombosis occurred in one JAK2-V617F positive patient. Two major vascular events were reported in the MPL-W515L positive JAK2-V617F negative woman (myocardial and mesenteric infarction).

Cytogenetic analyses rarely revealed rare karyotypic abnormalities including trisomy 8 and 21. Cytoreductive treatment consisted of hydroxyurea (n=9), hydroxyurea and anagrelide (n=1), hydroxyurea, anagrelide and cyclophosphamide (n=1), hydroxyurea and melphalan (n=1) and melphalan (n=1). One patient received interferon-α. In the JAK2-V617F, negative group, 9 patients died. The causes of death were unknown (n=2), pneumonia (n=1), stroke (n=2), myocardial infarction (n=1), lung cancer (n=1), and acute leukemic transformation (n=2). One patient with RARST-BP disease showed phenotypic features of acute myelomonocytic leukemia and acute erythroleukemia. The other patient who transformed to acute myelomonocytic leukemia with normal karyotype had been treated with hydroxyurea for 9 years, while no therapy prior to the onset of AML was given in the other patient. In the JAK2-V617F positive group 2 patient died (gall bladder adenocarcinoma: n=1; unknown cause: n=1).