

Summary:

Introduction: Circulating endothelial progenitor cells (EPC) may provide an endogenous repair mechanism to counteract ongoing endothelial damage. Antineutrophil cytoplasmic antibody - associated vasculitis (AAV) is an inflammatory disorder of small- to medium-sized vessels with relapsing/remitting progression and endothelial injury is a major feature of AAV. EPC thus may play an important role in the pathogenesis of AAV, or serve as a useful marker for monitoring and/or prediction of outcomes in patients with AAV.

Hypotheses: EPC number in patients with AAV could be altered. The decreased capacity for endothelial regeneration paralleled by low EPC numbers could increase the risk of relapse in patients with AAV.

Patients and methods: We have measured EPC in healthy volunteers, patients with AAV, chronic kidney failure (CKD) and atherosclerosis by a colony-forming assay. We have investigated the relation between the numbers of EPC, clinical and laboratory characteristics of the patients, and long-term outcomes of patients with AAV.

Results: Patients with AAV had a significantly lower number of EPC than healthy subjects, but not than patients with CKD or atherosclerosis. The cumulative relapse-free survival increased stepwise across three increasing baseline levels of endothelial progenitor cells.

Conclusion: Patients with AAV have a significant and persistent deficiency of circulating EPCs. A low number of EPCs could reflect an impaired mechanism of vascular repair and may contribute to repeated relapses in these patients.