

patient's marrow is a mosaic of mutant and nonmutant cells, and the peripheral blood always contains both GPI-negative (PNH) cells and GPI-positive (non-PNH) cells: in most cases the former prevail.

Thrombosis is one of the most immediately life-threatening complications of PNH, and yet one of the least understood in its pathogenesis. It could be that deficiency of CD59 on the PNH platelet causes inappropriate platelet activation; however, other mechanisms are possible. In very rare cases PNH can be caused by biallelic mutations of the *PIGT* gene, in the absence of a *PIGA* mutation. In these cases, because GPI is produced but cannot bind to proteins, the clinical picture is further complicated by the coexistence of a chronic inflammatory state.