

Abstract

Background:

Hemophilia is an inherited bleeding disorder caused primarily by a deficiency of coagulation factors. Patients with hemophilia (PWH) may have reduced cardiovascular disease (CVD) due to hypocoagulability. We conducted a systematic review to investigate the extent of CVD and atherosclerosis in PWH and associated risk factors.

Methods:

A systematic literature search was conducted of the following databases from inception through June 1, 2022: Pubmed, Embase, Ichushi, CINAHL and the Cochrane Library. No restrictions were applied regarding publication date or language, and case-control studies of PWH assessing CVD were included. We assessed the methodological quality using the Joanna Briggs Institute (JBI) critical appraisal tool.

Results:

We included five case-control studies. Methodological quality was judged as low risk in one study, moderate risk in three studies, and high risk in one study. In three studies, the cardiac risk factors were similar in cases and controls, but in one study the HDL-cholesterol levels were significantly lower in PWH. Intima media thickness (IMT), which morphologically indicates the degree of atherosclerosis, was significantly thinner in PWH in one study, and minimally reduced in another. There were no differences in the degree of coronary artery stenosis between PWH and controls in one study, but in another study, flow-mediated dilatation (FMD) was significantly decreased in moderate-severe and mild hemophiliacs compared with controls.

Conclusion:

PWH may have similar cardiovascular risks as non-hemophiliacs. However, it is still

unclear whether PWH are protected from atherosclerosis because results on the degree of atherosclerosis are conflicting. Further research is necessary to investigate the CVD risk in PWH.

Keywords: Hemophilia A, hemophilia B, CVD, atherosclerosis