A systematic review of mortality statistics and causes of death in people with congenital haemophilia (PwCHA)

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Summary

A current evidence-based understanding of mortality in congenital haemophilia A (HA) is absent.

A total of 17 records from 2010 to 2020 reporting mortality and/or cause of death in people with haemophilia (PwH) were analyzed.

Bleeding, human immunodeficiency virus (HIV), hepatitis C virus (HCV), and liver diseases are leading causes of death among PwH.

Reporting of cause of death was highly diverse and the methodology used is varied. A unified approach is needed to understand mortality in people with congenital haemophilia (PwHA).

Published mortality ratios suggest a raised mortality risk for PwH.

- As a ratio, the risk of death in PwH compared with that of the general population ranged from 0.86 (standardized mortality ratio) to 2.2 (hazard ratio) in all PwH,4,14 and from 2.4 (standardized mortality ratio) to 6.6 (hazard ratio) in people with severe haemophilia.14,15
- These ranges suggest a raised mortality risk, particularly in severe haemophilia (Figure 2).

Records reporting causes of death in PwH (15) were highly diverse.

- Records varied in size, population (age, comorbidities), location, and time.10–16,19–24 Inconsistent reporting of long-term outcomes limits evidence on mortality in PwH.
- The number of deaths reported in a single record ranged from 12 to 784.
- Differences in patient management and available treatments across high- and low-income populations met the criteria for this systematic review.

Differences in patient management and available treatments across high- and low-income populations included in the systematic review.

Primary cause of death changed over time.

- There were numerous departures in categorization and reporting of causes of death; nonetheless, broad trends were consistently observed across different records (Figure 4).
- Deaths relating to HIV and hepatitis B have been generally decreasing since the 1990s.14,15
- Cardiovascular disease is an increasingly prevalent cause of death in PwH, as improved treatment and prophylaxis has increased lifespan expectancy.14,15

Conclusions

Decreasing mortality rates in PwH were observed in recent decades. While limits to mortality and other reports and studies have been compiled, it remains limited by the publication mortality ratios suggest there is still an excess risk of death in PwH compared with the general population, particularly in severe haemophilia.

The categorization of death in the literature was highly diverse, limiting understanding of mortality in haemophilia.

A unified approach to reporting mortality and cause of death is needed to understand mortality in PwH and to monitor changes as treatments continue to progress.

References


Acknowledgments

Disclosure: Financial support was received by Shaile Shraddha K., Shahla Maleki, and Christine C. Brod for the preparation of this manuscript. The authors have no conflicts of interest.

Presented at the National Hemophilia Foundation (NHF) Bleeding Disorders Virtual Conference | August 1–8, 2020