Intracranial hemorrhage in hemophiliacs: data from the national cohort, FranceCoag Network.

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Hemophilia provides mainly hematoma and hemarthrosis, but intracranial hemorrhages (ICH) are responsible for a significant morbidity and mortality: mortality has been decreased by replacement therapy, but neurologic sequelae may be serious.

This work is a descriptive analysis of ICH in the hemophiliac population registered in the national cohort FranceCoag. The data are those available from 1994 to 2011, describing the type and severity of hemophilia, the number of ICH, the age of onset and incidence according to the age, the number of deaths, viral infections, prophylactic treatment and inhibitor's detection.

The population includes 5520 patients with the following distribution of hemophilia: 83% type A and 17% type B; 37% severe forms, 21% moderate and 42% mild. 131 ICH were reported in 118 patients (2.1%) and 10% of these patients had more than one episode. The ICH occur more often in severe hemophilia (72%) but is also observed in moderate (21%) and minor (6%) forms. 23 patients died (19.5%). ICH can occur at any age but especially in the first 2 weeks of life (14% of cases). The incidence is the highest in this early neonatal period (930.5/1000 patient-year (PY)) and it decreases with age, to remain steady after the age of 2 years (2/1000PY).

Viral infections (HIV and hepatitis C) are significantly more frequent in patients with ICH, and 11% of the patients had an inhibitor at the onset of ICH.

After 1 month old, ICH are mostly spontaneous and traumas are reported in only 40% of cases.

In conclusion, the prevalence of ICH in this population is the same as described in studies published in the past and more recently. Moreover, FranceCoag Registry provides interesting epidemiological knowledge. Two major risk factors of ICH are underlined: the severity of hemophilia and the neonatal period.