Haemophilia Registry of the Medical Committee of the Swiss Haemophilia Society

Update and annual survey 2008

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Summary

The Swiss Haemophilia Registry of the Medical Committee of the Swiss Haemophilia Society was established in 2000. Primarily it bears epidemiological and basic clinical data (incidence, type and severity of the disease, age groups, centres, mortality). Two thirds of the questions of the WFH Global Survey can be answered, especially those concerning use of concentrates (global, per capita) and treatment modalities (on-demand versus prophylactic regimens). Moreover, the registry is an important tool for quality control of the haemophilia treatment centres. There are no informations about infectious diseases like hepatitis or HIV, due to non-anonymisation of the data. We plan to incorporate the results of the mutation analysis in the future.

Keywords

Haemophilia, registry, treatment modality, factor use, quality control

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The base for the IT concept of the Swiss Hemophilia Registry was set in the year 2000. In 2004, the registry was transformed to the actual internet based form with double password security. The latest records from September 30th 2008, are presented here.

Patients

A total number of 856 patients (compared to 839 in year 2007) were included in the registry, corresponding to an increase of 2%. 51 patients were not assigned to a specific center and, because of lack of updating, excluded for the evaluation.

Type of haemophilia and other rare coagulation disorders

Generally speaking, the distribution of the registered coagulopathies has not changed over the last years as shown in Figure 1: haemophilia A 525 (65%), haemophilia B 106 (13%), von Willebrand disease 118 (15%), other coagulopathies 56 (7%), including deficiencies concerning:

- afibrinogenaemia (n = 9),
- factor VII (n = 23),
- factor X (n = 3),
- factor XI (n = 3),
- factor XIII (n = 14),
- factor V/VIII (n = 4).

Disease severity was classified according to the ISTH standards (1): severe haemophilia A/B < 1%, moderate 1–5%, and mild 6–40%. The corresponding results are shown in Figure 2: severe type A patients 188 (39%), moderate type A 100 (21%), mild type A 194 (40%), severe type B patients 26 (28%), moderate type B 36 (39%), and mild type B 31 (33%). Data were missing in 56 patients.

In 2008, there were only two newborns with newly diagnosed haemophilia A. Age distribution of the patients remained stable over the years with a current average age of 37 years.

The current inhibitor status was positive in 19 patients with haemophilia A and one patient with haemophilia B. High responding inhibitors were present in 13 patients (12 with haemophilia A and 1 haemophilia B) and low responding inhibitors in 7 patients, all with haemophilia A. However, the numbers were not fully representative, as data were missing in 95 patients.

Data on treatment types are shown in Figure 3: In Switzerland overall, on-demand treatment still exceeds prophylaxis for both haemophilia A and B. In haemophilia A...
346 (66%) patients performed on-demand treatment vs. 102 (19%) with a prophylactic treatment. The figure in haemophilia B was even more obvious: 78 (74%) on-demand vs. 13 (12%) prophylactic treatment. Over time, however, a progressive shift was observed towards prophylactic treatment, also in adults (data not shown).

Recently, a new treatment group (sporadic therapy) (2), representing 8% of haemophilia A and 9% of haemophilia B patients, was introduced to describe mainly mild haemophiliacs requiring such a minimal treatment that it cannot be called on-demand. It is highly probable that not all concerned patients have already been correctly transferred from the on-demand to the sporadic group. Treatment status of about 5 to 6% of all patients remained unknown.

**Mortality**

Death registration represents a small file in the registry. In the period 1996–2008, a total number of 58 patients died. During the year 2008 six patients died: three from cancer, one each from bleeding and liver disease and one from an unknown cause. The last HIV-related death was back in 2003.

**Factor concentrate**

The module includes following components:
- name and type of the concentrate,
- delivery of the concentrate: amount in U or mg, date of delivery and treatment indication
  - home treatment,
  - acute bleeding,
  - surgery,
  - stockpile for holidays,
- total amount of concentrate use per year,
- history of the actual concentrate and former ones.

The main data input comes from an automatic file transfer from the pharmaceutical companies which deliver the concentrates for home treatment to the registry. The total amounts of delivered concentrates are reported quarterly to the administrator responsible for the data input.

**Quality control**

Currently there are 47 authorized users in the registry, all accredited members of the Medical Committee of the Swiss Hemophilia Society. The history of their logins as well as the quality and the regularity of their inputs in the registry are controlled once a year. Compliance to the rules set by the Medical Committee is one of the key criteria for the accreditation and re-accreditation/validation of the corresponding haemophilia reference or treatment centers.

Currently, two reference and eight treatment centers for children and five reference and six treatment centers for adults are established in Switzerland. Apart from three small centers, all actually meet the compliance...
criteria, yearly editing the medical data of at least 80% of their patients and reporting on the follow-up of at least 3 (for treatment centers) or 20 (for reference centers) severe haemophiliacs during the last 12 months.

The registry will start to exert a continuous control of the last check-up dates; this will be a very valuable tool to prospectively assess quality of medical care in haemophilia and help to ensure at least one annual check-up in an accredited center for patients with severe disease. Currently, compliance rate to this rule is only about 50%.

**Conclusions**

The current status and the outlook of the registry are very encouraging. After 14 years it really starts to be self-acting. For haemophilia specialists it has become a motivating tool to put data in, as it provides useful epidemiological, clinical but also economical data.

The registry soon will be used for prospective quality control of haemophilia centers and care in Switzerland.

The data of the Swiss Registry will enable international comparisons in the fields of clinical and epidemiological research in haemophilia (4). It is planned to expand the spectrum of registered diagnoses, newly including severe inherited platelet disorders (e.g. Bernard-Soulier syndrome and Glanzmann’s thrombasthenia) and other coagulopathies.

Finally and after obtaining every individual informed consent to that, the registry will integrate the results from the national mutation analysis of haemophilia families in Switzerland.

Patients indirectly take advantage from the registry, as they get accurate and up-to-date information on accredited haemophilia centers in the country. Swiss medical authorities as well as suppliers show increasing interest in the data of the registry for their planning of emergency supplies of factor concentrates and prediction of overall usage in Switzerland. The registry will require continuous efforts of the Medical Committee and support from the Swiss Hemophilia Society Board to meet highest quality requirements.

**Interessenkonflikt**

Alle Autoren erklären, dass kein Interessenkonflikt besteht.

**References**