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.

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:
- afibrinogenemia (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 current inhibitor status was positive in 75 patients (18%): responding inhibitors in 45 patients, not responding in 30. 9% of the haemophilia A patients showed inhibitors in 8 patients and 1 haemophilia B.

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.

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

Zusammenfassung

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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.

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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.

Data can also be put in manually by any user. Despite the convenient and easy handling of the module, data are probably not completely collected yet.

In 2004 the software was enhanced by a statistics program allowing evaluation of the concentrates in various combinations of selection, quantity and period of time. The different factor concentrates used are presented in Figure 4.

In Switzerland as in other countries (3), recombinant products clearly lead the market in haemophilia A with about 75% vs. 25% plasma-derived factor VIII. In haemophilia B, plasma-derived factor IX concentrates remain clearly on top: 92% vs. 8% rFIX. It is somewhat disappointing to note that the type of concentrate remains unknown in many patients (30–40%). The total amounts of concentrates used between October 2007 and September 2008 are shown in Figure 5.

**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...
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 in data, 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**