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The Swiss Paediatric Surveillance Unit (SPSU)

Data on rare diseases in Switzerland are scarce. Such diseases may cause high mortality or severe morbidity and can therefore be of public health importance. An established system for the monitoring of such diseases did not exist in our country. In a small country like Switzerland this gap can only be filled by a nationwide collaboration of clinicians and researchers. The Swiss Paediatric Surveillance Unit (SPSU) is an active surveillance system for rare paediatric disorders or rare complications of more frequent diseases in hospitalized children. The surveillance unit is a common project of the Swiss Paediatric Association (SPA) and the Swiss Federal Office of Public Health (SFOPH). It has been developed in analogy of the British system (British Paediatric Surveillance Unit, BPSU)[1, 2] and is managed by an advisory committee which currently consists of five representatives of the two beforementioned organisations (chairman: PD Dr. G. Schubiger, Lucerne). The main duty of the committee is to judge the implementation of study protocols submitted by potential study investigators. The administration of the SPSU is provided by the SFOPH.

The main objective of the unit is to provide a surveillance system for diseases of potential public health importance. With rare diseases, only data from the whole country (or even several countries) allow the collection of a sufficient number of cases (Table 1). The system will also allow a quick response to problems arising by new or emerging disorders or interventions (e.g. the detection and monitoring of rare adverse vaccine reactions). In special situations, short term surveillance of more common diseases would also be possible. Another objective is the coordination of research. The unit provides an active nationwide surveillance system to paediatric researchers. The system is simple, flexible and minimises paperwork. Furthermore it aims to lessen the burden of reporting physicians which is related to requests for information from many different sources. If needed, the unit will provide advice and assistance to researchers. Finally, it will increase the awareness of less common disorders.

- Surveillance of rare disorders or rare complications of more common diseases in hospitalized children
- Quick response to emerging problems
- Coordination of research
- Increase awareness of less common disorders

Table 1. *SPSU objectives.*

Current performance

At present, all 39 Swiss paediatric training units participate in the surveillance system. The responsible physicians at each clinic receive an anonymous reporting card on a monthly basis on which they only have to report the number of cases observed during the last month at their clinic. It is important that the card is also returned if no diseases under surveillance are observed. In cases of non-response, one to two reminders are mailed to the responsible persons to guarantee a high level of completeness. At the beginning, all participants received a description of the surveillance system as well as short study protocols for each disease under surveillance (including objectives, case definitions and reporting instructions). Case reports are immediately transmitted to the concerned researcher (study investigator). He himself then contacts the reporting physi-

cian who usually has to complete a short anonymous questionnaire of not more than two pages. All further work, including additional investigations, data analyses and publication is done by the study investigator. The use of this surveillance system is free of charge in the first two years, afterwards a contribution will usually be requested from study investigators. Regular annual reports will be published in the SFOPH Bulletin and sent to all participants. Study investigators have to provide an annual overview on their study subject for this purpose.

The initial surveillance programme covers four diseases: vitamin K deficiency bleeding (VKDB), congenital rubella (CR), congenital toxoplasmosis (CT) and acute flaccid paralysis (AFP). The VKDB surveillance aims to evaluate morbidity and mortality of the disease after the introduction of a new oral vitamin K formulation. The surveillance of CR is part of the monitoring of the vaccination programme aimed at the elimination of rubella in Switzerland. With an actual rubella vaccination coverage of about 80% [3], an increase in congenital cases cannot be excluded in the future. In such a situation, the SPSU should make it possible to react promptly. The CT surveillance aims to provide missing data on disease occurrence and severity in order to formulate adequate recommendations for prenatal screening. The surveillance of AFP is part of the WHO poliomyelitis eradication programme; for proof of polio elimination in our country, a reliable surveillance system is needed. It must be assured that all cases of AFP are investigated for polio viruses. Underreporting by mandatory notifications may be important for polio, CR and CT, as has been demonstrated explicitly for other diseases (e.g. tetanus) which are notifiable by physicians[4]. Indeed, two of the last three polio cases, which were diagnosed in 1987, were only known to the SFOPH by pure chance and this was one and a half years after their occurrence.

The inclusion of new studies (up to a total of eight per year would be reasonable) will be judged on a regular basis by the advisory committee using the criteria listed in Table 2. Priority will be given to the surveillance of diseases of potential public health importance or if a possible impact can be expected. Interested researchers have to submit a short study protocol to the unit's chairman covering the most important features. For final inclusion, a complete protocol has to be provided according to the "Guidelines for study applications" (Richtlinien für die Aufnahme von Studien/Directives pour la soumission d'études). For more details please contact the SPSU secretary at the SFOPH (Ms Th. Kiener, Tel. 031/323 88 16) or the chairman (PD Dr. G. Schubiger, Tel. 041/25 31 51).

First results

Two parameters, which allow the evaluation of the effectiveness of the surveillance system are the number of reporting cards returned and the number of confirmed cases. In the first four months, the completeness of reporting was 100% for January and February (one mailing), 100% for March and 98% for April. Four provisional cases of AFP, three of CT, one of CR and one of VKDB were reported until the end of May. Additional investigations of these cases are ongoing. Based on data from the British Unit, the following number of confirmed or probable cases per year would be expected to be reported in Switzerland: AFP: 4–8, VKDB: 1–3, CT: 1–2, CR: 0–2. The small number of expected cases provides the possibility to include other diseases with slightly higher incidences in the future.

- Rarity of the condition
- Public health importance
- Need for rapid response
- Clear and practicable case definitions
- Methods of case ascertainment
- Useful and important research objectives
- Duration of surveillance
- Resources for study management
- Workload for paediatricians

Table 2. *Main criteria for the consideration of study protocols.*

Discussion

Paediatric surveillance units have proven to be useful tools in different countries. The longest experience comes from England, where the British Paediatric Association in collaboration with the Communicable Disease Surveillance Centre have run a surveillance system of this kind since 1981 (e.g. for Reye syndrome). The BPSU system became fully operational in 1986[1]. Two other countries in Europe, Holland and Germany, started surveillance programmes in 1992[5,6]. Table 3 gives a short overview of these three units. Other projects have been started in Australia (in 1993)[6] and in Malaysia (in 1994).

The reporting rate in all three European countries is 90% or more (Table 3). This is similar to Switzerland, and points to a good acceptance of these surveillance systems by the paediatricians. In England and Ireland, virtually all consultant paediatricians in clinical practices are now included. 28 studies have been completed so far (e.g. vitamin K deficiency bleeding, insulin dependent diabetes, congenital toxoplasmosis, acute rheumatic fever, MMR-associated meningoencephalitis or subacute sclerosing panencephalitis). An increasing number of publications

	UK	NL	D
Name	BPASU[1]	NSCK[4]	ESPED[7]
Institutions	BPA[2] (CDSC[3])	DPA[5] TNO[6]	GPA[8]
Start	1986	1992	1992
Participating paediatricians	1350	350	470
Reporting rate	90–94%	91–93%	90–94%
Completed studies	28	2	2
Conditions currently under surveillance (start)	9	8	9
	Reye Syndrome (1986)	Coeliac disease (1992)	Hib infections (1992)
	Aids/HIV (1990)	Acute flaccid paralysis (1992)	Diabetes mellitus (1992)
	Congenital rubella (1990)	Diabetes mellitus (1993)	Ondine's curse (1992)
	Hib vaccine failures (1992)	Neural tube defects (1993)	Vit. K def. bleeding (1993)
	Biliary atresia (1993)	Hib infections (1993)	Kawasaki syndrome (1993)
	Congenital syphilis (1993)	HIV/Aids (1995)	Pertussis complications (1993)
	MCAD[9] (1994)	Haemolytic disease of the newborn (1995)	Acute renal failure (1993)
	Adverse effects of labour and birth in water (1994)	Postneonatal mortality in premature born infants (1995)	Meningococcal infections (1994)
	Neonatal diabetes (1994)		Tick borne encephalitis (1994)

[1] British Paediatric Association Surveillance Unit (formerly BPSU), [2] British Paediatric Association, [3] Communicable Disease Surveillance Centre, [4] Nederlans Signalerings-Centrum Kindergeneeskunde, [5] Dutch Paediatric Association, [6] Instituut voor Praeventieve Gezondheidszorg TNO, [7] Erhebungsstelle für seltene pädiatrische Erkrankungen in Deutschland, [8] German Paediatric Association, [9] Medium chain acyl CoA dehydrogenase deficiency.

Table 3. *Paediatric surveillance units in Europe.*

have resulted from the BPSU surveys[7–18]. Based on the data from the congenital toxoplasmosis surveillance in 1989/90, a realistic estimate of 14 symptomatic cases per year (including 8 with CNS involvement) was made. The great disparity between these 14 cases and the estimation of 54 cases assuming a 2 per 1000 infection rate in pregnancy, a 40% transmission rate and a proportion of 10% severely symptomatic neonates, was one of the reasons not to recommend a screening programme[19]. Screening of acute toxoplasmosis in pregnancy was not judged to provide a proven benefit and should therefore not, at present, be offered routinely. There was even the possibility that such a programme might cause more harm than benefit. Some studies are done, or have been done, in more than one country, e.g. acute flaccid paralysis in England, Holland and Switzerland, diabetes in England, Holland and Germany, congenital rubella in England and Switzerland (in preparation in Holland). Some combined results of these common surveillance projects will be presented in September 1995 at the annual meeting of the European Society for Paediatric Research in Alicante, Spain. Also, a new application for EC funding for a concerted action programme will be undertaken this year in order to stimulate the development of similar units in other European countries.

Based on the experience of other countries, useful results can also be expected from the Swiss surveillance unit. The success of the SPSU depends upon the good collaboration of its participants and researchers, as well as on the support of the SPA and the SFOPH. We would like to thank all participants for the good start of this surveillance unit in our country.

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