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CORRESPONDENCE

Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis

Improving the Support to Victims

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Stevens-Johnson Syndrome (SJS) and toxic epidermal necrolysis (TEN) are severity variants of the same process of acute destruction of the epithelium of the skin and mucous membranes by a violent immune response that resembles, in many aspects, the acute rejection of a graft. The reaction is usually initiated by, and primarily directed against, drugs. Less frequently, it may be caused by infectious agents. The mortality rate is even higher than previously suspected and sequelae are nearly constant [1]. Less than 20 % of persons suffering from SJS/TEN survive without resulting harm for the rest of their lives. These figures completely support the denomination of 'victims' used by patient associations.

Dr Butt and co-authors [2] should be congratulated for their original approach in evaluating the impact of these terrible diseases on the daily life of survivors, using a systematic analysis of narratives of experiences posted on patient websites. One may suspect that patients who complain on the Internet are not representative. In my own experience, a substantial proportion of survivors suffering debilitating sequelae prefer 'turning the page' or 'trying to forget'.

In spite of this limitation, the findings of Dr Butt and co-authors [2] are strongly consistent with prior experience of patient associations, and my own 'expert' impression that survivors mainly complain of insufficient information. They consider themselves poorly informed on the causes of their disease, risk for their relatives, risk of recurrence and future use of medications. Another major concern is the difficulty to find an adequate medical support to alleviate sequelae that may affect so many organs (skin, eyes, mouth, lung, genitalia, digestive tract, etc.). These lesions are often poorly known to organ specialists because of the rarity of SJS/TEN. Furthermore, most physicians are afraid to prescribe any systemic medication.

Such a situation obviously needs improvement. Poor information is unfortunately largely related to absence of evidence. Recent advances on genetic factors can be helpful for only a fraction of patients. With the partial exception of ocular lesions, the prevalence, mechanism, progression and management of sequelae are still largely unknown.

What can we do? Some priorities are not difficult to list.

First, listen carefully to patient complaints, individually and through patient associations, when existing [3, 4]. Collaboration with patient associations helped us to better evaluate the psychological distress resulting from SJS/TEN, improve the management of pain [5], detect unexpected sequelae [6] and to elaborate leaflets to better inform patients and their relatives.

Second, we must improve medical knowledge on the mechanisms of sequelae. Clinical and basic research is a prerequisite to the development of preventive or curative measures. Because of the rarity of the disease such research should be based on networks of experts and close collaboration with patient associations.

The third objective will be to provide access to adequate care and also to compensation for all victims of these terrible reactions. For them, the 'benefit-risk balance' of the drug responsible has been infinitely negative. Do they have to pay the price of a hazardous accident while hundreds of thousands of other users gain a benefit?

Unfortunately, these objectives require costly research projects that, up to now, were rarely funded.

I consider that both pharmaceutical companies and regulatory agencies should be more implicated in

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supporting research on severe adverse drug reactions (ADRs). Marketing of new drugs is regulated, which is a serious hint that the responsibility is not only that of the firms but is also *de facto,* endorsed by the public authorities that deliver marketing authorizations. Until authorities, e.g. the US FDA and the European Medicines Agency, and drug companies contribute to organizing more research on ADRs, the benefit-risk balance will remain flawed by better evaluation of the benefits than the risks.

References

1. Bounoua M, Valeyrie-Allanore L, Sekula P, et al. Evaluation prospective des séquelles de syndrome de Stevens-Johnson et de nécrolyse épidermique toxique dans une cohorte européenne de 339 malades. Ann Dermatol Venereol. 2007;134:7S12.
2. Butt TF, Cox AR, Oyebode JR, et al. Internet accounts of serious adverse drug reactions: a study of experiences of Stevens-Johnson syndrome and toxic epidermal necrolysis. Drug Saf. 2012;35(12): 1159–70.
3. An example to us all. Prescrire Int. 2012;21:200.
4. Stevens-Johnson Syndrome Foundation [online]. Available from http://sjsupport.org/. Accessed 19 Nov 2012.
5. Valeyrie-Allanore L, Ingen-Housz-Oro S, Colin A, et al. Pain management in Stevens-Johnson syndrome, toxic epidermal necrolysis and other blistering diseases [in French]. Ann Dermatol Venereol. 2011;138:694–7.
6. Gaultier F, Rochefort J, Landru MM, et al. Severe and unrecognized dental abnormalities after drug-induced epidermal necrolysis. Arch Dermatol. 2009;145:1332–3.