

# Management of Autoimmune Bullous Diseases in France: A Nationwide Network of 30 Centers

Nicolas Meyer, MD<sup>a,b,\*</sup>, Carle Paul, MD, PhD<sup>b,c</sup>,  
Pascal Joly, MD, PhD<sup>d</sup>

## KEYWORDS

- Autoimmune bullous diseases • Care • Guidelines
- Reference center

## ORGANIZATION OF DERMATOLOGIC HEALTH CARE IN FRANCE

The management of autoimmune bullous disease (AIBD) in France has always been under the guidance of dermatologists. The health care system in France is characterized by a combination of complementary public and private structures. Most dermatologists in France, representing 90% of the 3400 dermatologists practicing in the country, are office-based physicians.<sup>1</sup> It was estimated that in the year 2000, 14 million outpatient consultations were performed by office-based dermatologists. Severe skin diseases as well as skin cancer, including melanoma and skin lymphoma, are mostly taken care of by hospital dermatologists. It has been estimated that inpatients represented more than 100,000 hospital stays during the same period.<sup>2</sup> Since 1958, the academic dermatology departments of the 31 university hospitals have been staffed by full-time hospital physicians. In the year 2000, there were 102

dermatology departments in France (including 51 in university hospitals and 61 in general hospitals), totaling a capacity of 1403 beds.<sup>3</sup> Most of the inpatient capacities are located in academic dermatology departments (in the year 2000, 77% of the 1403 beds of dermatology departments were located in university hospitals), although some nonacademic dermatology departments are very active. AIBDs represent an important group of diseases that are mostly managed by academic and nonacademic dermatology departments. Since 1985, a research group called the French Autoimmune Bullous Disease Group has been created within the French Dermatology Society (Société Française de Dermatologie [SFD]). The French Autoimmune Bullous Disease Group has promoted optimal care for AIBDs and organized clinical trials. Among the landmark studies promoted by the French Autoimmune Bullous Disease Group are the demonstration of the efficacy and safety of topical corticosteroids in bullous pemphigoid (BP),<sup>4</sup> the use of rituximab

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<sup>a</sup> UMR 1037-CRCT, 20 24 r Pont Saint Pierre, 31052 Toulouse Cedex 3, France

<sup>b</sup> Dermatology Department, Larrey Hospital, chemin de Pouvoirville, TSA30030, 31059 Toulouse Cedex 9, France

<sup>c</sup> Paul Sabatier-Toulouse III University, 118 route de Narbonne 31062 Toulouse Cedex 9, France

<sup>d</sup> Dermatology Department, INSERM U905, Rouen University Hospital, 1 Rue de Germont, 76031 Rouen, France

\* Corresponding author. Service de Dermatologie, Hôpital Larrey, 24 chemin de Pouvoirville TSA30030, 31059 Toulouse Cedex 9, France.

*E-mail address:* meyer.n@chu-toulouse.fr

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in pemphigus vulgaris,<sup>5</sup> and the identification of risk factors for BP.<sup>6</sup>

### AIBD: DEFINITION OF THE DISEASES

AIBDs are a group of rare, acquired disorders characterized by:

- The onset of vesiculobullous lesions on the skin or mucous membranes
- The alteration of cutaneous or mucous membrane components
- The presence of pathogenic autoantibodies targeting structural proteins of the desmosomes and the dermal-epidermal junction (binding of autoantibodies on these desmosomal and hemidesmosomal components disrupts these intercellular or dermal epidermal junctions).

AIBDs are BP, cicatricial pemphigoid, autoimmune pemphigus (AIP), epidermolysis bullosa acquisita, herpetiformis dermatitis, pemphigoid gestationis, and linear IgA dermatosis. AIBDs are separated in 2 groups:

- Intraepidermal AIBD: AIP
- Subepidermal AIBD: all other AIBDs.

Some AIBDs may be severe, especially recalcitrant or relapsing types, leading to treatment side effects and potential lethality.

### EPIDEMIOLOGY OF AIBDS IN FRANCE

Most cases of AIBD occur sporadically, without evidence of geographic or familial clustering. Little is known on the epidemiology of AIBD in France. In 1995, a multicentric prospective epidemiologic study was conducted in 3 university hospitals in France.<sup>7</sup> One hundred cases were evaluated over a period of 35 months. The investigators evaluated the overall standardized incidence of subepidermal AIBD to be 10.4 cases per year and per million inhabitants. The most frequent subepidermal AIBD was BP (standardized incidence: 7 cases per million inhabitants per year), which represented 70% of the cases of AIBD. The incidence of other subepidermal AIBD comprised between 0.17 and 0.26 case per year and per million inhabitants. A similar study performed 15 years later in the same regions in France found an incidence rate for BP of 21.7 cases per million persons per year, corresponding to a doubling of the incidence over a 15-year period. Aging of the population may at least in part account for this dramatic increase in disease incidence. The incidence rate of subepidermal AIBD was shown to increase dramatically with age, with up to 224,

329, and 507 cases per million persons per year in the populations aged 75, 80, and 85 years or older, respectively. These results are close to those recently reported by Langan and colleagues<sup>8</sup> in the United Kingdom.

Among AIBDs, the epidemiology of AIP has been extensively studied (reviewed in Ref.<sup>9</sup>). There are 2 basic types of pemphigus: pemphigus vulgaris and pemphigus foliaceus. Although paraneoplastic and drug-induced pemphigus has also been described, its incidence is low. Only sporadic types of pemphigus foliaceus and pemphigus vulgaris have been described in France in contrast to other Mediterranean countries. The incidence of the various types of pemphigus varies from 1 country to another. Sporadic pemphigus vulgaris is the most common in Europe and in the United States, whereas endemic pemphigus foliaceus is more prevalent in Northern Africa, Turkey, and Southern America. Two epidemiologic studies on AIP were conducted in France.<sup>10,11</sup> The first study was conducted in 1995 in the Paris region, the most populated area of northwestern France, and evaluated the incidence of AIP to be 1.7 cases per year and per million inhabitants.<sup>10</sup> The second study was conducted over the 2002 to 2006 period in the region Midi-Pyrénées in the southwestern part of the country. The study included only dermatologist-validated cases of the disease and estimated the incidence of AIP at 1.55 cases per year and per million inhabitants.<sup>11</sup>

In western Europe, 4 studies were conducted on the epidemiology of AIBD between 2000 and 2010.<sup>8,11–13</sup> In a prospective study conducted in 2001 to 2002, Marazza and colleagues<sup>12</sup> evaluated the incidence of BP and AIP in Switzerland. A total of 168 patients were identified. The BP and AIP standardized incidence was estimated at 12.1 and 0.6 cases per year and per million inhabitants, respectively. Similar results were reported by Bertram and colleagues<sup>13</sup> in Germany (Lower Franconia): The most frequent AIBD was BP (standardized incidence = 13.4 cases per year and per million inhabitants). The third study<sup>11</sup> was aimed at evaluating the incidence of AIP in southwestern France, and provided no data on the incidence of other AIBD, and the fourth was registry-based and conducted in the UK by Langan and colleagues.<sup>8</sup> In the study by Langan and colleagues the incidences of BP and pemphigus vulgaris were estimated to be 4.3 cases per year and per 100,000 inhabitants and 0.7 case per year and per 100,000 inhabitants, respectively.

Despite the limited number of studies available, all recent data on the epidemiology of AIBD support that AIBD are rare diseases and that the most frequent of them is BP.

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