Panorama of Autoimmune and Autoinflammatory Diseases in Internal Medicine at the University Hospital Center (UHC) of the Point G

Soukho Assétou Kaya1,2, Keïta Kaly1*, Sy Djibril1,2, Traoré Djénébou1,2, Dembélé Ibrahim Amadou1, Mallé Mamadou1, Cissoko Mamadou1, Sangaré Barry Boubacar Oumar1, Keïta Adramé1, Keïta Mamadou1, Nyanké Romuald1, Diarra Bacary1, Cissé Idrissa Ahmadou2,3, Diakité Mahamadou2,4,5, Dembélé Mamadou6, Traoré Abdel Kader6, Traoré Hamar Alassane6

1Department of Internal Medicine at the University Hospital Center of the Point G, Bamako, Mali
2Faculty of Medicine and Odontostomatology (FMOS), University of Sciences, Techniques and Technologies of Bamako (USSTB), Bamako, Mali
3Department of Rheumatology at the University Hospital Center of the Point G, Bamako, Mali
4University of Clinical Research Center (UCRC) at the University Hospital Center of the Point G, Bamako, Mali
5Malaria Research and Training Center (MRTC), University of Sciences, Techniques and Technologies of Bamako (USSTB), Bamako, Mali
6Faculty of Medicine, Kankou Moussa University (UKM), Bamako, Mali
Email: *keitakaly@gmail.com, hopitalpointg@hotmail.com

Abstract

Introduction: Panorama studies of autoimmune and auto-inflammatory diseases are still very little carried out in Africa and particularly in Mali. The objective of this descriptive study with retrospective collection was to describe the epidemiological and clinical profile of all autoimmune and auto-inflammatory diseases in the department of internal medicine at the University Hospital Center of the Point G. Methods: This was a descriptive study with a retrospective survey of the records of patients hospitalized for autoimmune and auto-inflammatory diseases in the department of internal medicine at the CHU of Point G for a study period of 15 years from January 1, 2005 to December 31, 2019. We included in the study all patients hospitalized for autoimmune and auto-inflammatory diseases. Results: During the study period (January 31, 2005 to December 31, 2019), 6383 patients were hospitalized in internal medicine at the University Hospital Center of the Point G, of which 317 patients presented with autoimmune and/or auto-inflammatory disease with an average annual hospital recruitment rate of 21 ± 7.87 cases per year. The female sex accounted for 64.98% with a sex ratio of
0.54. The mean age of patients was 35.27 ± 16.27 years and the extreme ages were 07 and 79 years. Out of the 317 medical records included according to our inclusion criteria, there were 07 cases of association between autoimmune disease and autoinflammatory disease, i.e. 14 cases of autoimmune and autoinflammatory diseases. A total of 331 autoimmune diseases and/or auto-inflammatory diseases were collected, i.e. a frequency of 5.19%, including 291 cases of autoimmune diseases (221 cases of organ-specific autoimmune diseases and 70 cases of systemic autoimmune diseases) and 40 cases of autoinflammatory diseases (no case of monogenic forms, 08 cases of “systemic” polygenic forms and 32 cases of “organ-specific” polygenic forms). Organ-specific autoimmune diseases were dominated by type 1 diabetes (141 cases), Graves’ disease (48 cases) and systemic autoimmune diseases by systemic lupus erythematosus (43 cases), rheumatoid arthritis (16 cases). Among the auto-inflammatory diseases, the “systemic” polygenic forms were dominated by Horton’s disease (02 cases) and the “organ-specific” polygenic forms by gout (16 cases), ulcerative colitis (08 cases).

**Conclusion:** It appears from our study that autoimmune and autoinflammatory diseases are characterized in internal medicine by their frequent occurrence in women and preferably between 25 and 44 years of age with very disparate distribution. We also observed a predominance of organ-specific autoimmune diseases over systemic ones, and “organ-specific” polygenic autoinflammatory diseases over “systemic” ones.

**Keywords**

Autoimmune Disease, Autoinflammatory Disease, Internal Medicine, Mali

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1. **Introduction**

Autoimmune diseases are defined as all pathological manifestations linked to the involvement of effectors of the immune system, B lymphocytes and T lymphocytes, specific to the antigens of the organism to which this system belongs (self-antigens) [1]. They are very heterogeneous and are usually classified into two groups: organ-specific autoimmune diseases and systemic autoimmune diseases [2].

Systemic autoimmune diseases are diseases in which the target antigen is distributed in different organs or tissues of the body [2]. These are mainly connective tissue diseases and autoimmune vascularitis. The hospital frequency of connective tissue diseases ranged from 0.20% to 1.60% [3] [4] [5] depending on the studies. The most frequent connective tissue diseases in the study done by Télessou et al. were systemic lupus erythematosus followed by scleroderma and rheumatoid arthritis [6], whereas in the study done by Dioussé et al. they were represented by systemic lupus erythematosus followed by scleroderma and dermatomyositis [3].

Organ-specific autoimmune diseases are diseases in which the target antigen is located in a tissue or a cell [2], including autoimmune type 1 diabetes, autoimmune thyroiditis (Graves’ disease, Hashimoto’s thyroiditis, and De Quer...
The prevalence of organ-specific autoimmune diseases was 1.15% according to a Nigerian study, the most representatives of which was Graves’ disease followed by autoimmune thrombocytopenic purpura [7].

As for auto-inflammatory diseases, they are due to an abnormality of innate immunity. There are no elevated or pathogenic autoantibodies and no activated T-lymphocytes, as opposed to autoimmune diseases. They are subdivided into two groups: monogenic autoinflammatory diseases and polygenic autoinflammatory diseases [8] [9].

Monogenic auto-inflammatory diseases are a small number of afflictions that share common clinical features. The mutation is carried by a gene of innate immunity in this specific case [8] [9], such as familial mediterranean fever, cryopyrin-associated auto-inflammatory disease, Blau syndrome [8] [9]. The prevalence of familial mediterranean fever ranged from 1 - 5 cases per 10,000 inhabitants [10]. It should be noted that monogenic auto-inflammatory diseases are exceptional in sub-Saharan Africa but cases are commonly reported in North Africa [11].

Clinical phenotype of the polygenic auto-inflammatory diseases may be very different from these hereditary fevers, notably due to the absence of intermittent symptoms. In this case, several genes are involved in the dysregulation of innate immunity [8] [9]. In order to facilitate understanding, according to their typical or usual clinical form [12]-[17], they can also be subdivided into two groups: “systemic” polygenic auto-inflammatory diseases and “organ-specific” polygenic auto-inflammatory diseases.

“Systemic” polygenic auto-inflammatory diseases are characterized by systemic impairment in their typical clinical form, such as sarcoidosis, non-autoimmune vascularitis (Behçet’s disease, Horton’s disease), Still’s disease. In the Senegalese series in 2019, these were represented by order of frequency: Behçet’s disease, Still’s disease, sarcoidosis, and atrophic polychondritis [18].

“organ-specific” polygenic auto-inflammatory diseases are also characterized by specific organ impairment in their usual clinical form, such as spondyloarthropathies, microcrystalline arthropathies, chronic inflammatory bowel diseases. Spondyloarthropathies followed by gout, pseudo-rhizomelic polyarthritis were the most frequently encountered polygenic auto-inflammatory diseases in the Senegalese series [18].

The objective of this study with retrospective data collection was to describe the epidemiological and clinical aspects of all autoimmune and auto-inflammatory diseases in the Department of internal medicine at the University Hospital Center of the Point G.

2. Methods

This was a descriptive study with retrospective data collection for 15 years (January 1, 2005 to December 31, 2019). It was carried out in the internal medicine Department at the University Hospital Center of the Point G in Bamako. We included in this study all the medical records of patients hospitalized for autoimm-
mune and/or autoinflammatory disease during the study period. Patients seen for autoimmune and auto-inflammatory diseases in the internal medicine department at the University Hospital Center of the Point G in the outpatient consultation and patients hospitalized for autoimmune and auto-inflammatory diseases in the internal medicine department at the University Hospital Center of the Point G outside the study period were not included in this study. This was an exhaustive sampling of all cases of hospitalization for autoimmune and/or auto-inflammatory diseases during the study period. Diagnosis for autoimmune and autoinflammatory diseases was established on the basis of clinical and paraclinical data and/or validated diagnostic criteria according to the type of autoimmune and autoinflammatory diseases. The data were collected on the pre-established survey form, including: epidemiological (age, sex, profession, residence) and clinical (reason of hospitalization, discharge diagnosis) characteristics. Data entry and analysis were done using SPSS version 22 software. Quantitative data were presented as mean and standard deviation if the distribution was normal, otherwise as median and interquartile range. Qualitative data were presented as numbers and percentages. In this work, no association or causality tests were performed. The hospitalization register was used (identification of patient records and determination of the total number of the study population) in strict confidentiality and was returned and filed in the archive room immediately after exploitation.

3. Results

During the study period (January 31, 2005 to December 31, 2019), 6383 patients were hospitalized in internal medicine at the University Hospital Center of the Point G of which 317 patients presented with autoimmune and/or auto-inflammatory disease with an average annual hospital recruitment rate of 21 ± 7.87 cases per year. The female sex accounted for 64.98% with a sex ratio of 0.54. The age range of 25 - 34 years represented 28.71% of the study population. The mean age of patients was 35.27 ± 16.27 years and the extreme ages were 07 and 79 years. Housewives were found in 37.22% of cases. Patients came from an urban zone in 64.04% of cases (Table 1). Endocrine manifestations were the reason for hospitalization in 23.78% of cases, followed by general manifestations in 15.33% of cases and digestive manifestations in 12.22% of cases (Figure 1).

Out of the 317 medical records included according to our inclusion criteria, there were 07 cases of association between autoimmune disease and autoinflammatory disease, i.e. 14 cases of autoimmune and autoinflammatory diseases. A total of 331 autoimmune diseases and/or auto-inflammatory diseases were collected, i.e. a frequency of 5.19%, including 291 cases of autoimmune diseases (221 cases of organ-specific autoimmune diseases and 70 cases of systemic autoimmune diseases) and 40 cases of autoinflammatory diseases (no case of monogenic forms, 08 cases of “systemic” polygenic forms and 32 cases of “organ-specific” polygenic forms) (Figure 2). Organ-specific autoimmune diseases were dominated by type 1 diabetes (141 cases), Graves’ disease (48 cases) (Table 2)
and systemic autoimmune diseases by systemic lupus erythematosus (43 cases), rheumatoid arthritis (16 cases) (Table 3). Among the autoinflammatory diseases, the “systemic” polygenic forms were dominated by systemic non autoimmune vascularitis (6 cases): Horton’s disease (02 cases), periarthritis nodosa (01 case), vascularitis of undetermined origin (1 case) followed by systemic sarcoidosis and Still’s disease (01 case each) (Table 4) and the “organ-specific” polygenic forms by chronic inflammatory rheumatism (n = 22 cases): gout (16 cases), reactive arthritis, psoriatic rheumatism, psoriatic rheumatism and pseudo rheumatoid arthritis (01 case, respectively) followed by chronic inflammatory bowel diseases (n = 10 cases): ulcerative colitis (08 cases), Crohn’s disease (2 cases) (Table 5).

Figure 1. Distribution of patients by reason for hospitalization.

Figure 2. Overall distribution of patients according to autoimmune and autoinflammatory diseases.
Table 1. Distribution of patients by sociodemographic data.

| Sociodemographic data | Number of cases (N=317) | Percentage |
|-----------------------|-------------------------|------------|
| **Gender**            |                         |            |
| Male                  | 111                     | 35.02      |
| Female                | 206                     | 64.98      |
| **Age group**         |                         |            |
| 05 - 14 years         | 20                      | 6.31       |
| 15 - 24 years         | 69                      | 21.77      |
| 25 - 34 years         | 91                      | 28.71      |
| 35 - 44 years         | 50                      | 15.77      |
| 45 - 54 years         | 31                      | 9.78       |
| 55 - 64 years         | 39                      | 12.30      |
| 65 - 74 years         | 14                      | 4.42       |
| ≥75 years             | 3                       | 0.95       |
| **Profession**        |                         |            |
| Housewife             | 118                     | 37.22      |
| Pupil/Student         | 49                      | 15.46      |
| Civil servant         | 36                      | 11.36      |
| Trader                | 28                      | 8.83       |
| Farmers*              | 24                      | 7.57       |
| Artisans              | 17                      | 5.36       |
| Worker                | 16                      | 5.05       |
| Not employed          | 4                       | 1.26       |
| Liberal profession    | 4                       | 1.26       |
| No information        | 21                      | 6.62       |
| **Residence**         |                         |            |
| Urban                 | 203                     | 64.04      |
| Rural                 | 76                      | 23.97      |
| Outside of Mali       | 11                      | 3.47       |
| No information        | 27                      | 8.52       |

Farmers*: Cultivator/Breeder/Fisherman.

Table 2. Distribution of patients by organ-specific autoimmune diseases.

| Organ-specific autoimmune diseases | Number | Percentage |
|-----------------------------------|--------|------------|
| **Type 1 diabetes**               | 140    | 63.35      |
| Grave’s disease                   | 47     | 21.27      |
| Autoimmune hemolytic anemia       | 10     | 4.52       |
| Biermer’s disease                 | 8      | 3.62       |
| Guillain Barré syndrome           | 7      | 3.17       |
| Autoimmune polyendocrinopathy     | 2      | 0.90       |
| Myasthenia gravis                 | 2      | 0.90       |
### Table 3. Distribution of patients by systemic autoimmune diseases.

| Systemic autoimmune diseases                                      | Number | Percentage |
|-------------------------------------------------------------------|--------|------------|
| Systemic lupus erythematosus                                      | 43     | 61.43      |
| Rheumatoid arthritis                                              | 16     | 22.86      |
| Sharp Syndrome/Mixed Connectivities tissue diseases (n = 6 cases; 8.57% of cases) | 4      | 5.71       |
| Systemic lupus erythematosus + rheumatoid arthritis + dermato-polymyositis + systemic sclerosis | 1      | 1.43       |
| Systemic lupus erythematosus + systemic scleroderma               | 4      | 5.71       |
| Systemic lupus erythematosus + rheumatoid arthritis               | 1      | 1.43       |
| Systemic lupus erythematosus + systemic sclerosis                 | 4      | 5.71       |
| Systemic lupus erythematosus + rheumatoid arthritis               | 1      | 1.43       |
| Systemic lupus erythematosus                                      | 4      | 5.71       |
| Dermato-polymyositis                                              | 1      | 1.43       |
| Systemic auto-immune vascularitis                                 | 0      | 0.00       |
| Total                                                             | 70     | 100.00     |

### Table 4. Distribution of patients by "systemic" polygenic autoinflammatory diseases.

| "Systemic" polygenic autoinflammatory diseases                      | Number | Percentage |
|-------------------------------------------------------------------|--------|------------|
| Systemic vascularitis non-autoimmune                              | 2      | 25.00      |
| Horton’s disease                                                  | 2      | 25.00      |
| Periarthritis nodosa                                               | 1      | 12.50      |
| Vasculitis of undetermined origin                                 | 1      | 12.50      |
| Autres                                                            | 1      | 12.50      |
|       Burger Angitis                                               | 1      | 12.50      |
|       Leukocytoclassical Vascularitis                              | 1      | 12.50      |
| Systemic Sarcoidosis                                              | 1      | 12.50      |
| Still’s disease                                                   | 1      | 12.50      |
| Total                                                             | 8      | 100.00     |

### Table 5. Distribution of patients by "organ-specific" polygenic autoinflammatory diseases.

| "Organ-specific" polygenic autoinflammatory diseases              | Number | Percentage |
|------------------------------------------------------------------|--------|------------|
| Microcrystalline arthropathies & Gout                           | 16     | 50.00      |
| Reactive arthritis                                               | 1      | 3.13       |
| Psoriatic rheumatism                                             | 1      | 3.13       |
| Ankylosing spondylitis                                           | 1      | 3.13       |
| Pseudo rheumatoid arthritis                                     | 1      | 3.13       |
| Others                                                           | 1      | 3.13       |
| Juvenile Idiopathic Arthritis                                   | 1      | 3.13       |
| Jaccoub’s Arthropathy                                           | 1      | 3.13       |
| Chronic inflammatory bowel diseases (n = 10 cases; 31.25% of cases) | 2      | 6.25       |
| Crohn’s disease                                                 | 2      | 6.25       |
| Ulcerative colitis                                               | 8      | 25.00      |
| Total                                                            | 32     | 100.00     |
4. Discussion

The lack of reliable hospital statistical data concerning an overview of these two major nosological entities (autoimmune diseases and autoinflammatory diseases) led us to undertake this work. The choice of internal medicine department is due to their multiple and varied activities.

As with any retrospective study, the interpretation of the results must take into account the pitfalls associated with the methodological approach of our study, which involved a certain number of biases: information bias (non-completeness of hospitalization records); selection bias (patients followed as outpatients in the internal medicine department, patients treated in other departments at the University Hospital Center of the Point G and other health structures); confusional bias (in particular the failure to perform certain specialized para-clinical examinations for the confirmatory diagnosis of certain autoimmune and auto-inflammatory diseases) and generalization bias (related to the hospital study site and monocentric recruitment). These biases may lead to under-estimate or over-estimate our study sample size. Notwithstanding these methodological shortcomings, we were able to comment on our results.

This was a descriptive study with a retrospective data collection during 15 years of activity (January 1, 2005 to December 31, 2019). It allowed us to apprehend the extent of the problematic of autoimmune and autoinflammatory diseases in internal medicine at the University Hospital Center of the Point G.

During the study period, 317 medical records were included according to our inclusion criteria with 331 autoimmune and auto-inflammatory diseases, i.e. a frequency of 5.19% with an average annual hospital recruitment rate of 21 ± 7.87 cases per year. The relatively low frequency of autoimmune and autoinflammatory diseases in our study could be explained by the economic and geographical inaccessibility of health care services by patients, but also by the use of traditional treatments, especially for chronic diseases [19]. Our result is consistent with the literature, as demonstrated by Sougué et al. in 2019, who reported 27 cases in 06 months of study in their study of chronic inflammatory rheumatism and autoimmune diseases [20]. In the department of dermatology in Burkina Faso, Konaté et al. collected 48 cases of systemic diseases over a study period from January 2017 to June 2019 [21].

Age group of 25 - 44 years represented 44.48% of the study population. The mean age of our patients was 35.27 ± 16.27 years and the extreme ages were 07 and 79 years. Our result is similar to those reported by Sougué et al. [20] and Konaté et al. [21] who found 47 and 38.4 ± 12.9 years respectively.

Out of the 317 patients included, there was a female predominance with 64.98%, a sex ratio of 0.54. This result contrasts with those of the authors from Burkina Faso [20] [21] who found a male predominance. This could be explained by the fact that our study was exhaustive. It included all autoimmune and autoinflammatory diseases.

In our study, our patients resided in urban areas in 64.04% of cases. A similar
finding was found in Burkina Faso by Konaté et al. [21].

Endocrine manifestations motivated hospitalization in 23.78% of cases followed by general manifestations with 15.33%. Contrary to our result, Sougué et al. [20] found osteoarticular manifestations to be the first reason for consultation. This difference can be explained by a different methodological approach between the two studies.

In our series, autoimmune diseases were found in 291 patients (4.56%) and accounted for 87.92% of the autoimmune and autoinflammatory diseases. In Niger, Garba et al. [7] reported a prevalence of autoimmune diseases of 7% in 11 month study in internal medicine. He noted a predominance of systemic autoimmune diseases with 89% of cases. This contrasts with those in our series, where organ-specific autoimmune disease accounted for 75.95% of cases. This difference could be explained by a shorter recruitment period in the Nigerian study. Our result is consistent with that of Denise et al. in the USA, who determined the prevalence of 24 systemic and organ-specific autoimmune diseases of which Graves’ disease and type 1 diabetes were the most prevalent, i.e. 1151.1/100,000 and 192/100,000 inhabitants respectively [22].

Connective tissue diseases in our study were found in 70 patients (1.10%) and those constituted 21.15% of the autoimmune and autoinflammatory diseases and 24.05% of the autoimmune diseases. Our result was comparable to that found by Zouna in Mali [5] who reported a frequency of 2.05%. However, it was higher than those found by Teclessou et al. [6] and Mijiyawa et al. [4] in Togo who found a frequency of 0.19% and 0.20% respectively. In Senegal, Dioussé et al. [3] reported a hospital prevalence of 0.29% over a 7-year study period at the department of dermatology. Panorally, in our series, systemic lupus erythematosus was the most frequent connective tissue disease with 63.35% of cases followed by rheumatoid arthritis with 21.27% of cases. This result is similar to those obtained by Teclessou et al. in Togo [6] (Systemic lupus erythematosus with 50.22% followed by rheumatoid arthritis with 21.64%), by Dioussé et al. in Senegal [3] (Systemic lupus erythematosus with 65.2% followed by Scleroderma with 21%). However, Mijiyawa et al. [4] in Togo and Zouna [5] in Mali found rheumatoid arthritis as the first connective tissue disease, respectively 29 cases and 77.78% of cases. This difference could be purely related to the methodological approach.

Organ-specific autoimmune diseases in the study population were noted in 221 patients (3.46%) and those represented 66.77% of the autoimmune and autoinflammatory diseases and 75.95% of the autoimmune diseases. Garba et al. in 2019 found a prevalence of 1.15% [7]. According to the panoramic profile, organ-specific autoimmune diseases were dominated in our study by type 1 diabetes with 63.35% of the cases followed by Graves’ disease with 21.27% of the cases. In Niger, Graves’ disease followed by immunologic thrombocytopenic purpura was the most frequent organ-specific autoimmune diseases [7]. The short recruitment period in this Nigerian study may explain these variable distributions of organ-specific autoimmune diseases.
In our study population, auto-inflammatory diseases were found in 40 patients with a hospital frequency of 0.63% of cases and constituted 12.08% of the autoimmune and autoinflammatory diseases. Some authors such as NZenze et al. [23] reported a prevalence of 2.34% in Gabon, Fall et al. [18] a prevalence of 8.1% in Senegal. This disparity between our results could be explained by the duration of the study and the recruitment sites, which differed in our respective studies. Fall et al. [18] reported one case of monogenic auto-inflammatory disease. However, in our series, no case of monogenic autoinflammatory diseases was found. Like our series, quasi-totality of the works on autoinflammatory diseases in sub-Saharan Africa remains dominated by the polygenic form [18] [20].

Among the patients with polygenic autoinflammatory diseases in our study, the “systemic” polygenic autoinflammatory diseases were recorded in 08 patients (0.13%) and represented 2.42% of the autoimmune and autoinflammatory diseases and 20.00% of the polygenic autoinflammatory forms. Systemic non-autoimmune vasculitis were the most frequent “systemic” polygenic auto-inflammatory diseases with 75.00% of cases. Systemic vasculitis was found with 4.9% of cases in the series of Fall et al. [18] in Senegal. Systemic sarcoidosis and Still’s disease were found in equal proportions in our study, which is 01 case each. As in our series, some authors such as Konaté et al. [21] and Sougué et al. [20] in Burkina Faso reported 01 case of systemic sarcoidosis in their study. In Senegal, Kane et al., reported 03 cases of Still’s disease in their series on systemic diseases [24].

“organ-specific” polygenic inflammatory diseases were noted in 32 patients (0, 50%) accounted for 9.67% of the autoimmune and autoinflammatory diseases and 80.00% of the polygenic autoinflammatory forms. Gout was the most common “organ-specific” polygenic autoinflammatory disease, accounting for 50.00% of cases in our study. The frequency observed in our study is comparable to that reported by Nzenze et al. [23] in Gabon who found that gout was the most frequent inflammatory arthropathy with 31.6% of cases. In contrast, Fall et al. [18] in Senegal and Divengi et al. [25] in the Democratic Republic of Congo (DRC) showed in their study that spondyloarthropathy was more frequent than gout, respectively 54.5% of cases versus 29% of cases and 57 patients versus 1 patient.

5. Conclusion

It appears from our study that autoimmune and autoinflammatory diseases are characterized in internal medicine by their frequent occurrence in women and preferably between 25 and 44 years of age with very disparate distribution. We also observed a predominance of organ-specific autoimmune diseases over systemic ones, and “organ-specific” polygenic autoinflammatory diseases over “systemic” ones.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.
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**Pre-established survey form**

Date of hospitalization: /__/__/ /__/__/ /__/__/__/
Date of discharged: /__/__/ /__/__/ /__/__/__/
Hospitalization duration (day): /__/__/__/
Medical record number: /__/__/__/__/
Telephone number: /__/__/ /__/__/ /__/__/

1. **Sociodemographic data**

| Gender          |   |
|-----------------|---|
| □ Male          |   |
| □ Female        |   |

| Age group       |   |
|-----------------|---|
| □ 05 - 14 years |   |
| □ 15 - 24 years |   |
| □ 25 - 34 years |   |
| □ 35 - 44 years |   |
| □ 45 - 54 years |   |
| □ 55 - 64 years |   |
| □ 65 - 74 years |   |
| □ >75 years     |   |
| □ No information|   |

| Profession      |   |
|-----------------|---|
| □ Housewife     |   |
| □ Pupil/Student |   |
| □ Trader        |   |
| □ Farmers (Cultivator/Breeder/Fisherman) |   |
| □ Worker        |   |
| □ Civil servant |   |
| □ Not employed  |   |
| □ Non information |   |
| □ Others        |   |
| □ Artisans      |   |
| □ Liberal profession |   |

| Residence       |   |
|-----------------|---|
| □ Urban         |   |
| □ Rural         |   |
| □ Outside of Mali |   |
| □ No information |   |
## 2. Clinical data

### 2.1. Reason for hospitalization

| Reason for hospitalization | 1: Yes | 2: No |
|----------------------------|--------|-------|
| General manifestations     |         |       |
| Prolonged fever            |         |       |
| Weight loss and/or asthenia and/or anorexia |         |       |
| Others                     |         |       |
| If other general manifestations (please specify): |         |       |

| Rhumatological manifestations |         |       |
|--------------------------------|--------|-------|
| Osteo-articular pain           |         |       |
| Joint deformity                |         |       |
| Joint swelling                 |         |       |
| Pathological fracture          |         |       |
| Others                         |         |       |
| If other rhumatological manifestations (please specify): |         |       |

| Dermatological manifestations |         |       |
|-------------------------------|--------|-------|
| Rashes                        |         |       |
| Pruritus                      |         |       |
| Alopecia                      |         |       |
| Edema of lower limbs and/or puffiness of the face |         |       |
| Others                        |         |       |
| If other dermatological manifestations (please specify): |         |       |

| Pleuro-pulmonary manifestations |         |       |
|---------------------------------|--------|-------|
| Cough                           |         |       |
| Dyspnea                         |         |       |
| Chest pain                      |         |       |
| Others                          |         |       |
| If other pleuro-pulmonary manifestations (please specify): |         |       |

| Digestive manifestations       |         |       |
|--------------------------------|--------|-------|
| Dysphagia                      |         |       |
| Nausea and/or vomiting         |         |       |
| Diarrhea                       |         |       |
| Abdominal pain                 |         |       |
| Abdominale distention          |         |       |
| Abdominal mass                 |         |       |
| Others                         |         |       |
| If other digestive manifestations (please specify): |         |       |
## Continued

**Endocrine manifestations**

- Polyuropolidipsia
- Ketoacidosis syndrome with or without coma
- Hyperthyroidism syndrome
- Others

If other endocrine manifestations (please specify):

**Neurological manifestations**

- Headache
- Disturbance of consciousness
- Convulsion
- Others

If other neurological manifestations (please specify):

**Other manifestations**

If other manifestations (please specify):

### 2.2. Diagnosis retained at discharge

| Diagnosis retained at discharge | 1: Yes | 2: No |
|---------------------------------|-------|-------|
| Autoimmune diseases             |       |       |
| Autoinflammatoires diseases     |       |       |
| Association between autoimmune diseases and autoinflammatoires diseases |       |       |

If association between autoimmune diseases and autoinflammatoires diseases (please specify):

#### 2.2.1. Autoimmune diseases

#### 2.2.1.1. Systemic autoimmune diseases

#### 2.2.1.1.1. Connective tissue diseases

| Connective tissue diseases | 1: Yes | 2: No |
|----------------------------|-------|-------|
| Systemic lupus erythematosus |       |       |
| Systemic scleroderma        |       |       |
| Dermato-polymyositis        |       |       |
| Rheumatoid arthritis        |       |       |
| Gougerot Sjogren syndrome   |       |       |
| Sharp Syndrome/Mixed Connectivities tissue diseases |       |       |
| Others                      |       |       |

If other (please specify):

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### 2.2.1.1.2. Systemic auto-immune vascularitis

| Systemic auto-immune vascularitis | 1: Yes | 2: No |
|-----------------------------------|--------|-------|
| Microscopic polyangiitis          | /............/ |
| Granulomatosis with polyangiitis (Wegener’s) | /............/ |
| Eosinophilic granulomatosis with polyangiitis (Churg and Strauss) | /............/ |

### 2.2.1.1.3. Association connective tissue diseases and systemic autoimmune vascularitis

| Association connective tissue diseases and systemic autoimmune vascularitis | 1: Yes | 2: No |
|--------------------------------------------------------------------------------|--------|-------|
| If association connective tissue diseases and systemic autoimmune vascularitis (please specify): | /............/ |

### 2.2.1.2. Organ-specific autoimmune diseases

| Organ-specific autoimmune diseases | 1: Yes | 2: No |
|-----------------------------------|--------|-------|
| Type 1 diabetes                   | /............/ |
| Autoimmune polyendocrinopathy    | /............/ |
| If Autoimmune polyendocrinopathy (please specify): | /............/ |
| De Quervain’s thyroiditis         | /............/ |
| Hashimoto’s thyroiditis           | /............/ |
| Postpartum thyroiditis            | /............/ |
| Grave’s disease                   | /............/ |
| Autoimmune adrenal insufficiency  | /............/ |
| Guillain Barré syndrome           | /............/ |
| Multiple sclerosis                | /............/ |
| Myasthenia                        | /............/ |
| Discoid lupus                     | /............/ |
| Psoriasis                         | /............/ |
| Pemphigus                         | /............/ |
| Bullous pemphigoid                | /............/ |
| Autoimmune vitiligo               | /............/ |
| Celiac disease                    | /............/ |
| autoimmune hepatitis              | /............/ |
| Autoimmune hemolytic anemia       | /............/ |
| Immunological thrombocytopenic purpura | /............/ |
| Biermer’s disease                  | /............/ |
| Others                             | /............/ |
| If other (please specify):        | /............/ |
2.2.1.3. Association systemic autoimmune diseases and organ-specific autoimmune diseases

| Association systemic autoimmune diseases and organ-specific autoimmune diseases |
|-----------------------------------------------------------------------------|
| If association systemic autoimmune diseases and organ-specific autoimmune diseases (please specify):…………………………...|

2.2.2. Autoinflammatory diseases

2.2.2.1. Monogenic autoinflammatory diseases

| Monogenic autoinflammatory diseases | 1: Yes | 2: No |
|------------------------------------|--------|-------|
| Mevalonate kinase deficiency       |         |       |
| Familial mediterranean fever       |         |       |
| Others                             |         |       |
| If other (please specify):         |         |       |

2.2.2.2. Polygenic autoinflammatory diseases

2.2.2.2.1. “Systemic” polygenic autoinflammatory diseases

| “Systemic” polygenic autoinflammatory diseases | 1: Yes | 2: No |
|-----------------------------------------------|--------|-------|
| Horton’s disease                              |         |       |
| Periartritis nodosa                           |         |       |
| Bechet’s disease                              |         |       |
| Rhumatoid purpura                             |         |       |
| Infectious angitis                            |         |       |
| Connectivite tissue diseases associated vascularitis |         |       |
| Drug-induced angitis                          |         |       |
| Vascularitis of undetermined origin           |         |       |
| Others                                        |         |       |
| If other (please specify):                    |         |       |
| Amyloidosis                                   |         |       |
| Systemic sarcoidosis                          |         |       |
| Still’s Disease                               |         |       |
| Others have the name of the other disease    |         |       |
| If other (please specify):                    |         |       |
2.2.2.2.2. “Organ-specific” polygenic autoinflammatory diseases

| “Organ-specific” polygenic autoinflammatory diseases | 1: Yes | 2: No |
|-----------------------------------------------------|--------|-------|
| Pseudo rheumatoid arthritis                          |        |       |
| Microcrystalline arthropathies                        |        |       |
| If microcrystalline arthropathies (please specify):  |        |       |
| Spondylarthropathies                                 |        |       |
| If spondylarthropathies (please specify):             |        |       |
| Others chronic inflammatory rheumatism               |        |       |
| If other chronic inflammatory rheumatism (please specify): |        |       |
| Chronic inflammatory bowel diseases                   |        |       |
| Crohn’s disease                                      |        |       |
| Ulcerative colitis                                   |        |       |
| Others                                               |        |       |
| If other (please specify):                           |        |       |

2.2.2.2.3. Association “systemic” polygenic autoinflammatory diseases and “organ-specific” polygenic autoinflammatory diseases

| Association “systemic” polygenic autoinflammatory diseases and “organ-specific” polygenic autoinflammatory diseases | 1: Yes | 2: No |
|----------------------------------------------------------------------------------------------------------------|--------|-------|
|                                                                                                                 |        |       |
| If association “systemic” polygenic autoinflammatory diseases and “organ-specific” polygenic autoinflammatory diseases (please specify):                        |        |       |

2.2.2.3. Association monogenic autoinflammatory diseases and polygenic autoinflammatory diseases

| Association monogenic autoinflammatory diseases and polygenic autoinflammatory diseases | 1: Yes | 2: No |
|--------------------------------------------------------------------------------------------|--------|-------|
|                                                                                             |        |       |
| If association monogenic autoinflammatory diseases and polygenic autoinflammatory diseases (please specify):                        |        |       |