



## The ESSDAI Explained: **Assessing Sjögren's Disease Activity**



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“**A clear understanding  
of disease activity  
helps us see where  
patients are—and  
where they're going.**”

— Audrey Gibson, PA-C, MSPAS, CCD

**OBJECTIVE:** Understand what ESSDAI measures and how it captures systemic disease activity in Sjögren's disease, how it reflects the disease's heterogeneous, systemic nature, and how APPs can apply ESSDAI-based questioning to uncover systemic (*sicca plus*) manifestations in patients who may be appropriate candidates for biologic therapy.

## **ESSDAI is the gold standard in clinical trials for assessing disease activity across 12 domains**

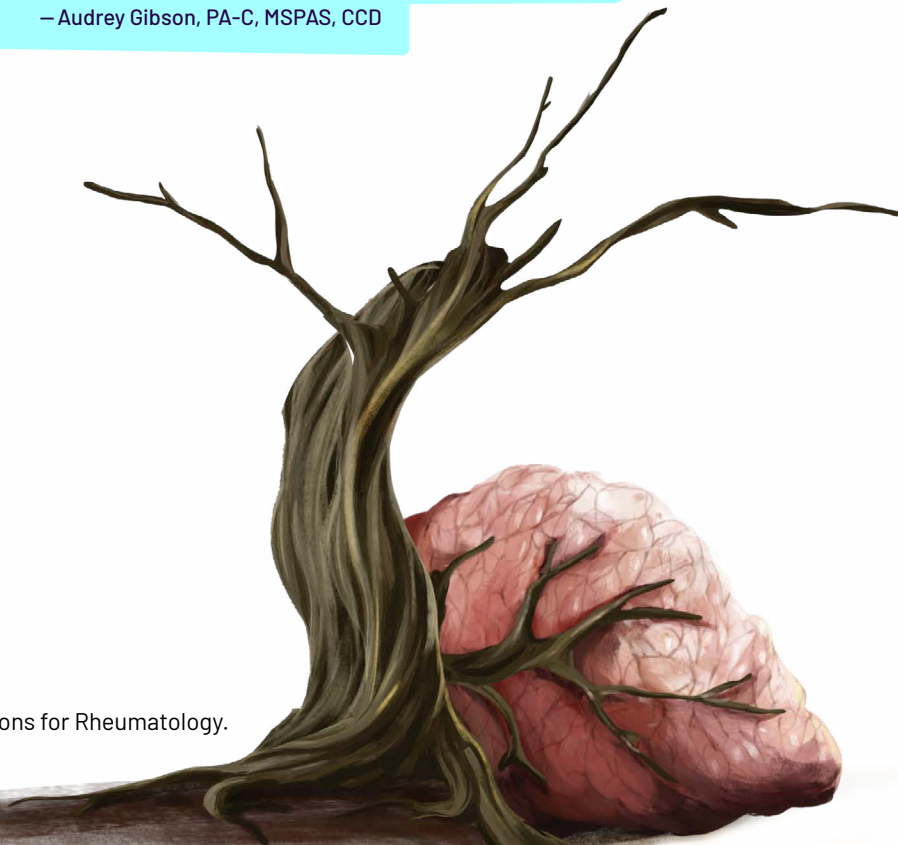
Sjögren's disease is a serious, systemic autoimmune disease that can be progressive and affect multiple organs.<sup>1-7</sup> Sjögren's represents a constant disease burden, with daily symptoms that significantly impact quality of life.<sup>8-10</sup>

Given this complexity and risk, guided clinician questioning can support a more structured approach to disease assessment, help uncover systemic manifestations, and enable longitudinal tracking of disease activity and treatment response. One tool is the **EULAR Sjögren's syndrome disease activity index (ESSDAI), which evaluates systemic disease activity across 12 organ domains.**<sup>11</sup> Developed and validated by experts, the ESSDAI is considered the gold standard end point to measure disease activity in clinical trials for Sjögren's disease. With clearly defined criteria to characterize activity level in each domain, the ESSDAI serves as an effective method for standardized and consistent patient evaluation.<sup>11</sup>

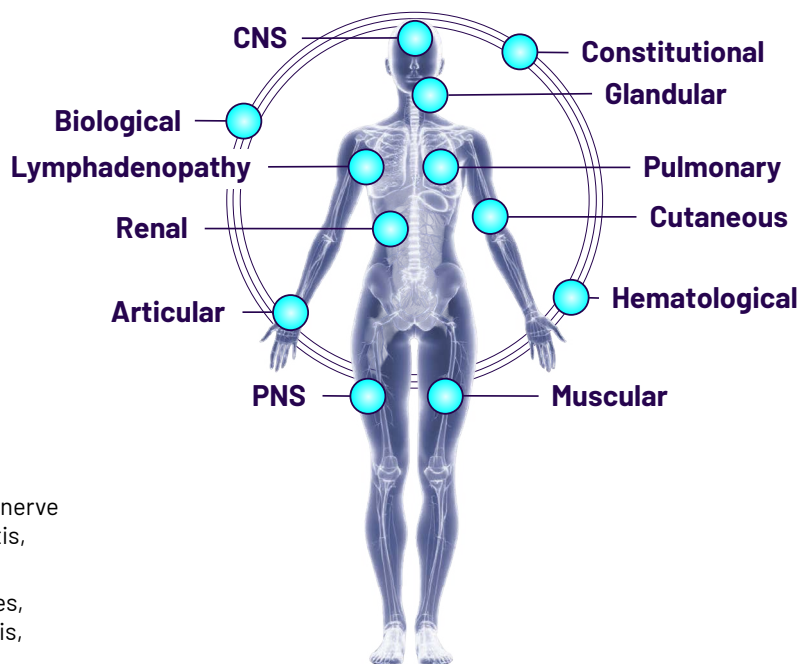
Appreciating the heterogeneous, systemic nature of Sjögren's disease is increasingly important as new therapies emerge. Familiarity with how systemic disease activity is characterized can help identify patients who may benefit from biologic therapies and support interpretation of clinical trial data in Sjögren's disease.

**“ESSDAI can serve as a reminder for comprehensive evaluation of all potentially involved systemic domains.”**

— Audrey Gibson, PA-C, MSPAS, CCD



## Assessing Systemic Involvement in Sjögren's: The 12 ESSDAI Domains



### CNS

- Moderately active CNS features, such as cranial nerve involvement, optic neuritis, MS-like syndrome
- Highly active CNS features, such as cerebral vasculitis, TIA, or seizures

### Biological

- Clonal component
- Hypocomplementemia
- Hypergammaglobulinemia
- High or recent decrease of IgG levels

### Lymphadenopathy

- Present in any nodal or inguinal region
- Current malignant B-cell proliferative disorder

### Renal

- Tubular acidosis
- Glomerular involvement with proteinuria
- Extramembranous glomerulonephritis
- Renal failure

### Articular

- Arthralgias in hands, wrists, ankles, and feet; morning stiffness
- Synovitis

### PNS

- Mild active PNS involvement, such as pure sensory axonal polyneuropathy
- Moderately active CNS involvement, such as axonal sensory-motor neuropathy
- Highly active PNS involvement, such as peripheral nerve involvement due to vasculitis

### Muscular

- Myositis
- Muscle weakness
- Elevated creatine kinase

### Constitutional

- Fever
- Night sweats
- Involuntary weight loss

### Glandular

- Enlarged parotid
- Submandibular or lacrimal swelling

### Pulmonary

- Persistent cough due to bronchial involvement
- Shortness of breath
- Interstitial lung disease

### Cutaneous

- Erythema multiforme
- Cutaneous vasculitis

### Hematological

- Autoimmune cytopenia with
- Neutropenia
- And/or anemia
- And/or thrombocytopenia
- Or lymphopenia

**Figure 1.** Systemic manifestations of Sjögren's disease across ESSDAI domains.<sup>11</sup> Domains may include manifestations beyond the examples shown.

## From Domains to Scores: Quantifying Disease Activity With ESSDAI

To calculate the total ESSDAI score, each domain is independently evaluated and assigned an activity level. Weighted domain scores are then summed to generate the total score.<sup>12</sup> When determining activity within each domain, only clinical features attributable to Sjögren's disease are included, with exclusion of manifestations related to comorbid conditions. Irreversible damage is also excluded, as the ESSDAI is designed to assess current, potentially reversible disease activity rather than cumulative organ damage.<sup>11</sup>

**Each domain is categorized by activity level (no, low, moderate, or high) based on defined clinical criteria<sup>12</sup>:**

- **No activity = 0**
- **Low activity = 1**
- **Moderate activity = 2**
- **High activity = 3**

The activity level score for each domain is then multiplied by its respective domain weight.<sup>12</sup> The total ESSDAI score is calculated as the sum of all weighted domain scores. Based on the total score, patients are classified as having low, moderate, or high systemic disease activity.<sup>13</sup>

### ESSDAI Domains

Domain Weights: 1 2 3 4 5 6

|                                                                                                 |                                                                                                       |                                                                                                       |
|-------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------|
|  Muscular 6  |  Renal 5           |  Articular 2     |
|  CNS* 5      |  Lymphadenopathy 4 |  Glandular† 2    |
|  PNS 5       |  Constitutional† 3 |  Hematological 2 |
|  Pulmonary 5 |  Cutaneous 3       |  Biological† 1   |

### ESSDAI Scoring<sup>12</sup>

|    |     |      |          |     |      |     |           |
|----|-----|------|----------|-----|------|-----|-----------|
| <5 | Low | 5-13 | Moderate | ≥14 | High | 123 | Max Score |
|----|-----|------|----------|-----|------|-----|-----------|

\*Domain does not contain low activity level. †Domain does not contain high activity level.

**While several ESSDAI domains can be evaluated intuitively at the bedside, others necessitate supplemental information, including laboratory parameters or imaging findings.**

—Audrey Gibson, PA-C, MSPAS, CCD

## ESSDAI Domain Scoring Deep Dive and Questions to Consider Asking Your Patients



### Muscular Domain<sup>11</sup>

Have you experienced any muscle pain or weakness?

| Activity Level | Domain Weight | Domain Score | Description                                                                                                                                                 |
|----------------|---------------|--------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------|
| No             | 6             | 0            | Absence of currently active muscular involvement                                                                                                            |
| Low            | 6             | 6            | Mild active myositis shown by abnormal EMG, MRI, or biopsy with no weakness and creatine kinase ( $N \leq CK \leq 2N$ )                                     |
| Moderate       | 6             | 12           | Moderately active myositis proven by abnormal EMG, MRI, or biopsy with weakness (maximal deficit of 4/5), or elevated creatine kinase ( $2N < CK \leq 4N$ ) |
| High           | 6             | 18           | Highly active myositis shown by abnormal EMG, MRI, or biopsy with weakness (deficit $\leq 3/5$ ) or elevated creatine kinase ( $>4N$ )                      |

CK, creatine kinase; EMG, electromyogram; MRI, magnetic resonance imaging; N, normal limit.

In the presence of these symptoms, the ESSDAI scoring guidance requires objective confirmation—such as via EMG, MRI, or muscle biopsy—to assign an activity level.<sup>11</sup>

**Myositis symptoms are not as common and require a deliberate effort to assess to ensure they are not overlooked.**

—Audrey Gibson, PA-C, MSPAS, CCD



### CNS Domain<sup>11</sup>

Have you experienced any headaches, dizziness, or confusion?

| Activity Level | Domain Weight | Domain Score | Description                                                                                                                                                                                                                    |
|----------------|---------------|--------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| No             | 5             | 0            | Absence of currently active CNS involvement                                                                                                                                                                                    |
| Moderate       | 5             | 10           | Moderately active CNS features, such as cranial nerve involvement of central origin, optic neuritis, or multiple sclerosis-like syndrome with symptoms restricted to pure sensory impairment or proven cognitive impairment    |
| High           | 5             | 15           | Highly active CNS features, such as cerebral vasculitis with cerebrovascular accident or transient ischemic attack, seizures, transverse myelitis, lymphocytic meningitis, multiple sclerosis-like syndrome with motor deficit |

These symptoms may prompt further clinical evaluation to assess CNS involvement, which could include features such as sensory or cognitive impairment.



## PNS Domain<sup>11</sup>

Have you experienced any numbness, tingling, or burning sensations in your extremities?

| Activity Level | Domain Weight | Domain Score | Description                                                                                                                                                                                                                                                                               |
|----------------|---------------|--------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| No             | 5             | 0            | Absence of currently active PNS involvement                                                                                                                                                                                                                                               |
| Low            | 5             | 5            | Pure sensory axonal polyneuropathy shown by NCS or trigeminal (V) neuralgia                                                                                                                                                                                                               |
| Moderate       | 5             | 10           | Axonal sensory-motor neuropathy (maximal motor deficit of 4/5)<br>Pure sensory neuropathy with presence of cryoglobulinemic vasculitis, ganglionopathy with symptoms restricted to mild/moderate ataxia, cranial nerve involvement of peripheral origin (except trigeminal (V) neuralgia) |
| High           | 5             | 15           | Axonal sensory-motor neuropathy with motor deficit $\leq 3/5$ , peripheral nerve involvement due to vasculitis (mononeuritis), severe ataxia                                                                                                                                              |

NCS, nerve conduction study.

Further evaluation can be guided by the established clinical recommendations from the American College of Rheumatology.

**PNS involvement in Sjögren's has not always been a focus in rheumatology.**

[Click here](#) to view the new guidelines that provide clearer directions for identifying and assessing PNS manifestations.<sup>14</sup>

**You have to specifically ask about respiratory symptoms—otherwise they can easily go undetected.**

— Audrey Gibson, PA-C, MSPAS, CCD



## Pulmonary Domain<sup>11</sup>

Have you been experiencing shortness of breath or a persistent cough?

| Activity Level | Domain Weight | Domain Score | Description                                                                                                                                                                                                                                |
|----------------|---------------|--------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| No             | 5             | 0            | Absence of currently active pulmonary involvement                                                                                                                                                                                          |
| Low            | 5             | 5            | Persistent cough due to bronchial involvement with no radiographic abnormalities on radiography, or radiological or HRCT evidence of interstitial lung disease with no breathlessness and with normal lung function test                   |
| Moderate       | 5             | 10           | Moderately active pulmonary involvement, such as interstitial lung disease shown by HRCT with shortness of breath on exercise (NYHA II) or abnormal lung function tests restricted to $70\% > DL_{CO} \geq 40\%$ or $80\% > FVC \geq 60\%$ |
| High           | 5             | 15           | Highly active pulmonary involvement, such as interstitial lung disease shown by HRCT with shortness of breath at rest (NYHA III, IV) or with abnormal lung function tests: $DL_{CO} < 40\%$ or $FVC < 60\%$                                |

$DL_{CO}$ , carbon monoxide diffusing capacity; FVC, forced vital capacity; HRCT, high-resolution computed tomography; NYHA, New York Heart Association.

Up to 20% of patients with Sjögren's disease develop interstitial lung disease.<sup>14</sup>



### Renal Domain<sup>11</sup>

Have you noticed any changes in your urine color or volume?  
Have you experienced any swelling in your legs or feet?

| Activity Level | Domain Weight | Domain Score | Description                                                                                                                                                                                                                                                                                      |
|----------------|---------------|--------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| No             | 5             | 0            | Absence of currently active renal involvement                                                                                                                                                                                                                                                    |
| Low            | 5             | 5            | Tubular acidosis without renal failure or glomerular involvement with proteinuria (between 0.5 and 1 g/day) and without hematuria or renal failure (GFR $\geq$ 60 mL/min)                                                                                                                        |
| Moderate       | 5             | 10           | Tubular acidosis with renal failure (GFR <60 mL/min) or glomerular involvement with proteinuria between 1 and 1.5 g/d and without hematuria or renal failure (GFR $\geq$ 60 mL/min) or histological evidence of extramembranous glomerulonephritis or important interstitial lymphoid infiltrate |
| High           | 5             | 15           | Glomerular involvement with proteinuria >1.5 g/d, or hematuria or renal failure (GFR <60 mL/min), or histological evidence of proliferative glomerulonephritis or cryoglobulinemia-related renal involvement                                                                                     |

GFR, glomerular filtration rate.

These symptoms may prompt further clinical evaluation to assess renal involvement, which could include serious features, such as renal failure.

**At every visit, we're reassessing for organ involvement, paying particular attention to the ones that can impact mortality.**

— Audrey Gibson, PA-C, MSPAS, CCD

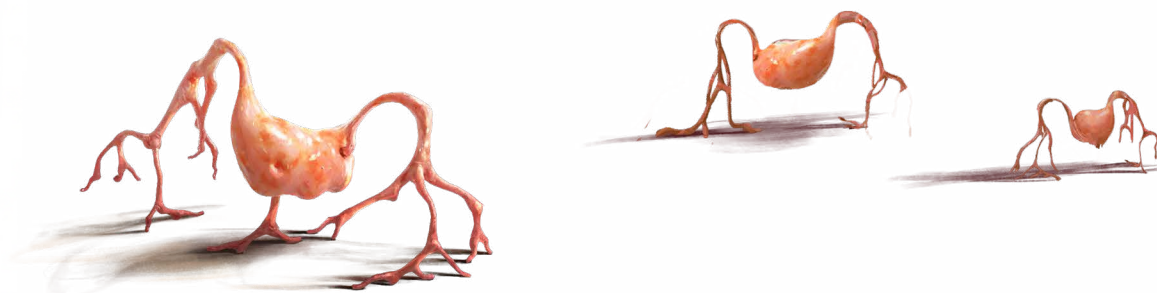


### Lymphadenopathy Domain<sup>11</sup>

Have you noticed any swollen lymph nodes?

| Activity Level | Domain Weight | Domain Score | Description                                                                                            |
|----------------|---------------|--------------|--------------------------------------------------------------------------------------------------------|
| No             | 4             | 0            | Absence of currently active lymphadenopathy involvement                                                |
| Low            | 4             | 4            | Lymphadenopathy $\geq$ 1 cm in any nodal region or $\geq$ 2 cm in inguinal region                      |
| Moderate       | 4             | 8            | Lymphadenopathy $\geq$ 2 cm in any nodal region or $\geq$ 3 cm in inguinal region, and/or splenomegaly |
| High           | 4             | 12           | Current malignant B-cell-proliferative disorder                                                        |

Patients with Sjögren's disease have a **10- to 44-fold increased risk of lymphoma** compared to healthy individuals.<sup>16</sup> Clinical features that have been associated with increased lymphoma risk include persistent salivary gland enlargement, cutaneous involvement, and lymphadenopathy.<sup>17,18</sup>





### Constitutional Domain<sup>11</sup>

Have you experienced fever or night sweats in the past 4 weeks?

| Activity Level | Domain Weight | Domain Score | Description                                                                                                     |
|----------------|---------------|--------------|-----------------------------------------------------------------------------------------------------------------|
| No             | 3             | 0            | Absence of currently active constitutional involvement                                                          |
| Low            | 3             | 3            | Mild or intermittent fever (99.5–101.3 °F)/night sweats and/or involuntary weight loss of 5%–10% of body weight |
| Moderate       | 3             | 6            | Severe fever (>101.3 °F)/night sweats and/or involuntary weight loss of >10% of body weight                     |

Constitutional symptoms reported are assessed in the context of overall disease activity and after exclusion of alternative causes, such as infection or other comorbid conditions.<sup>11</sup>



### Cutaneous Domain<sup>11</sup>

Have you noticed any changes in skin color or texture?  
Have you noticed any skin rashes or lesions?

| Activity Level | Domain Weight | Domain Score | Description                                                                                                                      |
|----------------|---------------|--------------|----------------------------------------------------------------------------------------------------------------------------------|
| No             | 3             | 0            | Absence of currently active cutaneous involvement                                                                                |
| Low            | 3             | 3            | Erythema multiforme                                                                                                              |
| Moderate       | 3             | 6            | Limited cutaneous vasculitis, including urticarial vasculitis, or purpura limited to feet and ankle, or subacute cutaneous lupus |
| High           | 3             | 9            | Diffuse cutaneous vasculitis, including urticarial vasculitis, or diffuse purpura, or ulcers related to vasculitis               |

Characterizing the type and extent of cutaneous involvement is important because skin involvement can be associated with increased risk of developing lymphoma.<sup>17</sup>



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**Figure 2.** Examples of cutaneous involvement seen in Sjögren's disease.



### Articular Domain<sup>11</sup>

Have you had pain in your hands, wrists, ankles, or feet in the past 4 weeks?  
Have you noticed any stiffness in the morning lasting for at least 30 minutes?

| Activity Level | Domain Weight | Domain Score | Description                                                                              |
|----------------|---------------|--------------|------------------------------------------------------------------------------------------|
| No             | 2             | 0            | Absence of currently active articular involvement                                        |
| Low            | 2             | 2            | Arthralgia in hands, wrists, ankles, and feet accompanied by morning stiffness (>30 min) |
| Moderate       | 2             | 4            | 1-5 (of 28 total count) synovitis                                                        |
| High           | 2             | 6            | ≥6 (of 28 total count) synovitis                                                         |

Clinical assessment of the articular domain focuses on identifying inflammatory features, including joint tenderness and swelling. In fact, in a study of 188 patients diagnosed with Sjögren's disease, the number of involved organs in patients with articular manifestations was found to be twice that of patients without articular manifestations, underscoring the importance of carefully tracking joint involvement.<sup>19</sup>



### Glandular Domain<sup>11</sup>

Have you noticed any swelling in your face or neck?

| Activity Level | Domain Weight | Domain Score | Description                                                                                                                              |
|----------------|---------------|--------------|------------------------------------------------------------------------------------------------------------------------------------------|
| No             | 2             | 0            | Absence of currently active glandular involvement                                                                                        |
| Low            | 2             | 2            | Small glandular swelling with enlarged parotid gland ( $\leq 3$ cm)<br>Submandibular ( $\leq 2$ cm) or lacrimal swelling ( $\leq 1$ cm)  |
| Moderate       | 2             | 4            | Major glandular swelling with enlarged parotid gland ( $> 3$ cm)<br>Important submandibular ( $> 2$ cm) or lacrimal swelling ( $> 1$ cm) |

Clinical examination can assess for enlargement of the major salivary glands, particularly the parotid and submandibular glands.



### Example of Glandular Swelling

Patient portrayal.



### Hematological Domain<sup>11</sup>

| Activity Level | Domain Weight | Domain Score | Description                                                                                                                                                                                                                                                                                                                                                                                                                                               |
|----------------|---------------|--------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| No             | 2             | 0            | Absence of currently active hematological involvement                                                                                                                                                                                                                                                                                                                                                                                                     |
| Low            | 2             | 2            | Cytopenia of autoimmune origin with <ul style="list-style-type: none"> <li>• Neutropenia (<math>1000 \leq \text{neutrophils} \leq 1500/\text{mm}^3</math>)</li> <li>• Anemia (<math>10 \leq \text{hemoglobin} \leq 12 \text{ g/dL}</math>) and/or</li> <li>• Thrombocytopenia (<math>100,000 \leq \text{platelets} \leq 150,000/\text{mm}^3</math>) OR</li> <li>• Lymphopenia (<math>500 \leq \text{lymphocytes} \leq 1000/\text{mm}^3</math>)</li> </ul> |
| Moderate       | 2             | 4            | Cytopenia of autoimmune origin with <ul style="list-style-type: none"> <li>• Neutropenia (<math>500 \leq \text{neutrophils} \leq 1000/\text{mm}^3</math>)</li> <li>• Anemia (<math>8 \leq \text{hemoglobin} \leq 10 \text{ g/dL}</math>) and/or</li> <li>• Thrombocytopenia (<math>50,000 \leq \text{platelets} \leq 100,000/\text{mm}^3</math>) OR</li> <li>• Lymphopenia (<math>\text{lymphocytes} \leq 500/\text{mm}^3</math>)</li> </ul>              |
| High           | 2             | 6            | Cytopenia of autoimmune origin with <ul style="list-style-type: none"> <li>• Neutropenia (<math>\text{neutrophils} &lt; 500/\text{mm}^3</math>)</li> <li>• Anemia (<math>\text{hemoglobin} &lt; 8 \text{ g/dL}</math>) and/or</li> <li>• Thrombocytopenia (<math>\text{platelets} &lt; 50,000/\text{mm}^3</math>)</li> </ul>                                                                                                                              |

In the context of the hematological domain, clinicians may assess for abnormalities such as cytopenias, including anemia, leukopenia, or thrombocytopenia, with laboratory evaluations to confirm hematologic involvement and to exclude alternative etiologies.



### Biological Domain<sup>11</sup>

| Activity Level | Domain Weight | Domain Score | Description                                                                                                                                                                                       |
|----------------|---------------|--------------|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| No             | 1             | 0            | Absence of currently active biological involvement                                                                                                                                                |
| Low            | 1             | 1            | Clonal component and/or hypocomplementemia (low C4 or C3 or CH50) and/or hypergammaglobulinemia or high IgG level between 16 and 20 g/L                                                           |
| Moderate       | 1             | 2            | Presence of cryoglobulinemia and/or hypergammaglobulinemia or high IgG level $> 20 \text{ g/L}$ , and/or recent onset hypogammaglobulinemia or recent decrease of IgG level ( $< 5 \text{ g/L}$ ) |

During assessment of the biological domain, clinicians may assess relevant laboratory parameters associated with systemic disease activity, including immunological and inflammatory markers. This could include abnormalities such as hypocomplementemia, hypergammaglobulinemia, or the presence of specific autoantibodies. For the purpose of the ESSDAI scoring, these findings are interpreted in the context of overall disease activity after exclusion of alternative causes.



## Applying ESSDAI: Patient Case Examples



### Nancy

Articular and  
Hematological Involvement

Not a real patient.

Nancy presents with significant inflammatory joint involvement, including synovitis affecting multiple joints (6) with both tenderness and swelling. This corresponds to **high activity in the articular domain**

$$\textcircled{3} \text{ high activity } \times \textcircled{2} \text{ domain weight } = 6$$

- Female, early 30s
- Severe mouth and eye dryness
- Fatigue

She also demonstrates hematologic involvement with cytopenia (absolute neutrophil count  $750 \text{ cells/mm}^3$ ), consistent with **moderate activity in the hematological domain**

$$\textcircled{2} \text{ moderate activity } \times \textcircled{2} \text{ domain weight } = 4$$

After applying the respective domain weights and summing the weighted scores, **Nancy's total ESSDAI score is**

$$6 + 4 = 10$$

indicating **moderate systemic disease activity**.



### June

Lymphadenopathy and  
Biological Involvement

Not a real patient.

June presents with lymphadenopathy, including swollen cervical lymph nodes ( $1.8 \times 2.2 \text{ cm}$ ), consistent with **moderate activity in the lymphadenopathy domain**

$$\textcircled{2} \text{ moderate activity } \times \textcircled{4} \text{ domain weight } = 8$$

- Female, late 50s
- Severe eye and mouth dryness
- Fatigue

She also demonstrates biological activity with low complement levels and elevated IgG level ( $19 \text{ g/L}$ ), corresponding to **low activity in the biological domain**

$$\textcircled{1} \text{ low activity } \times \textcircled{1} \text{ domain weight } = 1$$

After applying the respective domain weights and summing the weighted scores, **June's total ESSDAI score is**

$$8 + 1 = 9$$

indicating **moderate systemic disease activity**.

**Are you fully capturing systemic disease activity in your patients with Sjögren's?**

While the ESSDAI is primarily used in research settings, understanding the domains it captures can provide valuable perspective on how systemic manifestations relate to disease activity in Sjögren's disease, helping to inform clinical awareness and decision-making.<sup>11</sup>



**“A lot of patients have been dismissed—told it’s just dryness or just aging. Having a validated tool helps show patients that their disease is real and supports informed discussions about potential therapeutic strategies.”**

— Audrey Gibson, PA-C, MSPAS, CCD



[Click here to learn more practical tips on how to track disease progression](#)

**References:** 1. Brito-Zerón P et al. *Nat Rev Dis Primers*. 2016;2:16047. 2. Ramos-Casals M et al. *Ann Rheum Dis*. 2020;79(1):3-18. 3. Thalayasingam N et al. *Rheumatology (Oxford)*. 2021;60(suppl 6):vi53-vi61. 4. Kassan SS, Moutsopoulos HM. *Arch Intern Med*. 2004;164(12):1275-1284. 5. Vitali C et al. *Front Med (Lausanne)*. 2021;8:676885. 6. Negrini et al. *Clin Exp Med*. 2022;22(1):9-25. 7. Mariette X, Criswell A. *N Engl J Med*. 2018;378(10):931-939. 8. McCoy SS et al. *J Rheumatol*. 2021;48(7):1029-1036. 9. McCoy S et al. *Clin Rheumatol*. 2022;41:2071-2078. 10. Perella C et al. *Rheumatol Ther*. 2023;10:601-614. 11. Seror R et al. *RMD Open*. 2015;1(1):e000022. 12. Seror R et al. *Ann Rheum Dis*. 2010;69(6):1103-1109. 13. Seror R et al. *Ann Rheum Dis*. 2016;75(2):382-389. 14. Deboo A et al. *Arthritis Care Res*. 2026;0(0):1-15. 15. Flament T et al. *Eur Respir Rev*. 2016;25:110-123. 16. Retamozo S et al. *Lupus*. 2019;28(8):923-936. 17. Nocturne G, Mariette X. *Br J Haematol*. 2015;168:317-327. 18. Price EJ et al. *Rheumatology*. 2025;64:409-439. 19. Fauchais AL et al. *Rheumatology (Oxford)*. 2010;49(6):1164-1172.

