# Clinical characteristics of primary Sjögren's syndrome in adult patients diagnosed at age less than or equal to 35 years versus those over 35 years of age

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**Introduction:** Although there is little information, the debut of primary Sjögren's syndrome in adult patients aged less than or equal to 35 years, would be expressed with less pronounced Sicca manifestations and with a high degree of systemic commitment. For this reason, we decided to compare the behavior of the disease in the Argentine population with a diagnosis of primary Sjögren's syndrome at age less than or equal to 35 years versus those older than 35 years.

**Objective:** To describe and compare the systemic manifestations, assessed through the domains of the ESSDAI and the glandular manifestations, in Argentine adult patients diagnosed with primary Sjögren's Syndrome at age less than or equal to 35 years versus those over 35 years of age.

**Materials and Methods:** Cross-sectional, observational, analytical study. We analyzed the data of patients older than 18 years, with diagnosis of primary Sjögren's syndrome (American-European criteria 2002 / ACR EULAR 2016), included in the GESSAR database (Sjögren Syndrome Study Group of the Argentine Society of Rheumatology), with minus one control in the last 12 months. The presence of systemic manifestations assessed from the domains of the ESSDAI and the glandular manifestations, and as possible confounders were considered: sex, diabetes, hypothyroidism, treatments and time of follow-up in years of each group. The continuous variables were described as mean and standard deviation or median and interquartile range (RIQ), according to distribution and sample size. The categorical variables were expressed in percentages. For the bivariate analysis, for the continuous variables, the Student or Mann Whitney test was used, according to distribution and sample size. The categorical variables were analyzed using Chi square or Fisher's exact test, according to the expected frequency distribution table. A $p < 0.05$ was considered statistically significant.

**Results:** 665 patients were included. One hundred of them with an age at diagnosis ≤ 35 years and with a mean age at diagnosis of 29 ± 4 years, 92% of them women. The average age at diagnosis of the group > 35 years was 54 ± 11 years. Within the glandular manifestations, statistically significant differences were found between ≤ 35 years vs >35 years, in xerophthalmia (90.72% vs 95.64%, p: 0.04) and xeroderma (42.35% vs 57.36%, p: 0.03). No significant differences were found in xerostomía nor xerovagina. Statistically significant differences were found between ≤35 years vs >35 years in the following domains of ESSDAI: peripheral nervous system (4.05 vs 11.32, p: 0.03), respiratory (6% vs 15.40%, p: 0.01) and renal (6% vs. 1.59%, p: 0.02). In a subanalysis of ESSDAI domains, significant differences were found in arthritis (37.37% vs 27.16%, p: 0.04) and persistent dry cough (4.04% vs 11.47%, p: 0.03). No significant differences were found between ≤35 years vs >35 years in: follow-up time, diabetes, hipothyroidism, symptomatic treatment, treatment with pilocarpine, hydroxychloroquine, corticosteroids nor with immunosuppressants.

**Conclusion:** A significantly lower frequency of xerophthalmia and xeroderma was observed in the group ≤35 years old compared to those >35 years old. Regarding systemic activity, less involvement of the peripheral nervous system and pulmonary domain and higher in the renal domain, with statistically significant differences. These results suggest a lower glandular compromise in patients diagnosed at a younger age, without a characteristic differential pattern in terms of systemic involvement.

Table 1. General Characteristics.

| | Diagnosis ≤ 35 years n: 100 | Diagnosis > 35 years n: 565 | P value |
|---|---|---|---|
| Female sex (%) | 92.11 | 95.69 | 0.18 |
| Hypothyroidism ((%) | 15 | 21.95 | 0.115 |
| Diabetes (%) | 1 | 2.48 | 0.359 |
| Follow-up time (years) (mean, RIQ) | 4 (1-8) | 3 (1-6) | 0.15 |
| Corticosteroids (%) | 32.88 | 34.26 | 0.819 |
| Hydroxychloroquine (%) | 85.92 | 78.39 | 0.148 |
| Inmunosuppresants (%) | 15 | 17.20 | 0.588 |
| Symptomatic Treatm(%) | 90.22 | 93.03 | 0.565 |
| Pilocarpine (%) | 27.14 | 29.87 | 0.646 |

Table 2. Results.

| Glandular Manifestations | Diagnosis ≤ 35 years n: 100 (%) | Diagnosis > 35 years n: 565 (%) | P value |
|---|---|---|---|
| **Xerophthalmia** | **90.72** | **95.64** | **0.043** |
| Xerostomia | 87.37 | 90.33 | 0.377 |
| **Xeroderma** | **42.35** | **55.36** | **0.026** |
| Xerovagina | 30.86 | 39.18 | 0.153 |

| Systemic Manifestations (ESSDAI) | Diagnosis ≤ 35 years n: 100 (%) | Diagnosis > 35 years n: 565 (%) | P value |
|---|---|---|---|
| Articular | 66.00 | 68.32 | 0.647 |
| Cutaneous | 11.00 | 9.38 | 0.613 |
| **Pulmonary** | **6.00** | **15.40** | **0.012** |
| **Renal** | **6.00** | **1.59** | **0.016** |
| **PNS** | **4.04** | **11.31** | **0.015** |
| CNS | 1.00 | 2.65 | 0.279 |
| Haematological | 23.00 | 21.42 | 0.723 |
| Biological | 54.05 | 56.26 | 0.724 |
| Parotid swellig | 31.63 | 27.39 | 0.388 |
| Muscular | 2.02 | 1.62 | 0.513 |