

# Clinical Course of Segmental Vitiligo: A Retrospective Study of Eighty-Seven Patients

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**Background:** Vitiligo is an acquired disorder characterized by a progressive loss of melanocytes, which is difficult to manage and has an unknown prognosis. The subtype of segmental vitiligo (SV) has been established but it has not been adequately characterized. **Objective:** To collect long-term follow-up data for evaluating the clinical course of SV. **Methods:** This study included 87 patients who were diagnosed with SV and were monitored at a clinic. Patients were classified into the following three groups according to disease activity. **Results:** Among the patients with SV, 63.2% had stable disease, 14.9% had disease recurrence between two and four years after disease onset, and 21.8% had disease recurrence at four or more than four years after disease onset. Among the 44 patients (50.2%) who were monitored continuously over a four-year period, 19 (43.2%) experienced a recurrence at four or more than four years after disease onset. **Conclusion:** Our results suggest that, contrary to previous reports, some patients with SV may not experience disease stability over an extended period of time. Disease recurrence can occur after years of stability, and we propose that long-term follow-up data can be used to characterize SV. This information about the clinical course of SV has implications for treatment and prognosis. (*Ann Dermatol* 26(1) 61~65, 2014)

## -Keywords-

Prognosis, Recurrence, Segmental, Vitiligo

## INTRODUCTION

Vitiligo is an acquired disorder characterized by a progressive loss of melanocytes and a complex pathogenesis with a prevalence of approximately 0.5% worldwide. Although several theories have been proposed to explain the loss of epidermal melanocytes in this multi-factorial polygenic disorder, the precise cause remains unknown<sup>1</sup>. Patients with vitiligo present with one or more amelanotic macules that appear chalky or milky-white in color. Vitiligo is traditionally classified into either non-segmental or segmental vitiligo (NSV or SV, respectively)<sup>2,3</sup>. SV is characterized by macules in a linear or flag-like pattern of mosaicism with a dermatomal or quasi-dermatomal distribution<sup>1,4</sup>. This condition is not associated with autoimmune diseases and tends to have an early onset and more rapid progression compared to NSV<sup>4</sup>. Poliosis is also common in SV and is indicative of a poor response to phototherapy and standard medical treatments<sup>5,6</sup>. SV with leukotrichia usually requires surgical treatment, such as epidermal grafting<sup>6</sup>. However, there are a very few studies on the clinical course of SV. The purpose of this study was to evaluate the long-term disease activity of SV.

## MATERIALS AND METHODS

This retrospective study was conducted at a single tertiary medical center (Samsung Medical Center, Seoul, Korea). Eighty-seven Korean patients (41 males and 46 females) diagnosed with SV were treated and observed between March 1995 and September 2011. SV was defined as

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acquired leukoderma involving a unilateral cutaneous segment or delineated along the midline with a quasi-dermatomal distribution. Focal and mixed types of vitiligo were excluded. The follow-up period was defined as the time from the first to the last documented clinic visit. The disease duration was defined as the period between the initial disease onset and the first visit to our clinic. Data were collected from medical records and medical photographs. A thorough patient chart and photography review were used to determine the disease activity over the course of the follow-up period. Disease recurrence was defined as the appearance of new lesions or the spreading of preexisting lesions, and it was assessed in

each patient. Patients were then classified into one of the three groups based on the disease activity. In general, SV is thought to stabilize spontaneously by the second year of onset<sup>1-4</sup>, and therefore group 1 (typical rapid stabilization group) included patients who presented with stable disease or had active disease that had stabilized within two years of onset. We again used the two-year benchmark to determine the criteria for group 2, which included patients who experienced a recurrence between two and four years after the onset. Group 3 included patients who experienced disease recurrence at four or more than four years after the disease onset.



**Fig. 1.** Segmental vitiligo. Disease activity could be confirmed by a photo review. The black arrows in photos indicate sites of recurrence after a period of stability. (A, B) Male/13 years old (onset: at 8 years of age). The patient had white patches on the lower eyelid and perinasal area and the lesions progressed to the cheek after five years. (C, D) Male/19 years old (onset: at 12 years of age). The patient had a white patch on the left of neck and the lesions progressed to the right side after six years. (E, F) Female/16 years old (onset: at 4 years of age). She had been in a stable state for 10 years and visited the clinic for a recurrence at 15 years of age (E) and the lesions was progressing for one year (F). (G, H) Male/22 years old (onset: at 16 years of age). The patient had a white patch on the right side of the chin and the lesions progressed to the neck after four years. (I, J) Male/8 years old (onset: at 2 years of age). The patient had white patches on the inguinal area and the lesions progressed to the lateral side of thigh after five years.

## RESULTS

The mean age of patients with SV (n=87) at disease onset was 18.5 years (range: 2 to 63 years). Approximately half of the patients (n=44) presented to our clinic at more than 48 months after disease onset, while the average time of presentation was 38.7 months after onset. The mean follow-up period in Samsung Medical Center was 29.5 months (maximum: 150 months), and 23% of patients (n=20) were observed for 48 months or more.

After classifying the patients into three groups depending on the disease activity, the groups were analyzed. Patients in group 1 (n=55, 63.2%, 27 males, 28 females) presented with stable disease or had active disease that had stabilized within two years. In this group, the mean patient age at disease onset was 20.0 years (range: 2 to 63 years), the mean disease duration was 34.5 months (range: 1 to 636 months), and the average length of in-clinic follow-up was 20.9 months (maximum: 102 months). Patients in group 2 (n=13, 14.9%, 8 males, 5 females) showed a recurrence of disease between two and four years following onset. In this group, the average follow-up period was 29 months (maximum: 87 months). Patients in group 3 (n=19, 21.8%, 6 males, 13 females) experienced a recurrence at a minimum of four years after the disease onset and were followed-up for an average of 54.7 months (maximum: 150 months). The mean disease duration was 64.0 months (range: 1 to 360 months).

Among all of the groups, 57 patients (65.5%) demonstrated segmental facial involvement. Eighteen participants (20.7%) showed extensive involvement of two segments or the trunk. Among these 18 participants, half (n=9, 10.3%) had involvement of both the face and neck, and the remaining half (n=9, 10.3%) had vitiligo involving the trunk. Other sites involved included the neck only (n=4, 4.6%), the upper extremities (n=2, 2.3%) and the lower extremities (n=5, 5.7%). The long-term dormant group had a higher proportion of trunk and combined facial and neck involvement compared to the other groups (Fig. 1). Table 1 summarizes the clinical course of the studied 87 patients.

## DISCUSSION

It has been reported that SV responds poorly to medical treatment and stabilizes after an initial rapid spread. The etiopathogenesis of SV remains unclear; however, several hypotheses have been proposed, including neuronal mechanisms and a mosaicism hypothesis<sup>4,7</sup>. Ezzedine et al.<sup>8</sup> reported no statistically significant difference in sex or age at onset between patients with SV and NSV, while features of inflammation, autoimmunity and a familial background of vitiligo were strongly linked to NSV compared to SV in a prospective observational study. Mazereeuw-Hautier et al.<sup>9</sup> found that the percentage of patients with SV who experienced disease recurrence was

**Table 1.** Characteristics of patients with segmental vitiligo

Variable	Stable state within 2 years (group 1)	Active state between 2 and 4 years (group 2)	Active state after 4 years (group 3)	Total
Patients	55 (63.2)	13 (14.9)	19 (21.8)	87 (100)
Sex (male : female)	27 : 28	8 : 5	6 : 13	41 : 46
Age of onset (yr)	20.0±15.7, 2~63 (15.0, 10.0~30.0)	19.3±14.3, 2~42 (12.0, 7.0~17.0)	13.5±9.7, 2~40 (20.0, 7.0~35.0)	18.5±14.5, 2~63 (15.0, 8.0~22.0)
Duration of disease (mo)	34.5±89.9, 1~636 (18.0, 14.0~33.0)	17.0±13.2, 1~36 (42.0, 10.0~72.0)	64.0±83.1, 1~360 (12.0, 5.5~30.0)	37.9±81.9, 1~636 (18.0, 12.0~36.0)
Duration of observation (mo)	20.9±26.9, 0~102 (10.0, 2.0~36.0)	29.0±27.8, 0~87 (51.0, 22.0~81.0)	54.7±41.1, 0~150 (29.0, 0.5~41.5)	29.5±33.2, 0~150 (18.0, 4.0~48.0)
Time of recurrence (mo)		31.2±6.7 (24~47)	102.4±94.7 (48~480)	
Distribution	55 (100.0)	13 (100.0)	19 (100.0)	87 (100.0)
Face	40 (73.7)*	8 (61.5)*	9 (47.4)*	57 (65.5)*
Neck	3 (5.3)*	0 (0.0)*	1 (5.3)*	4 (4.6)*
Face & neck	5 (8.8)*	1 (7.7)*	3 (15.8)*	9 (10.3)*
Trunk	3 (5.3)*	2 (15.4)*	4 (21.1)*	9 (10.3)*
Trunk & face	0 (0.0)*	0 (0.0)*	1 (5.3)*	1 (1.1)*
Upper extremities	2 (3.5)*	0 (0.0)*	0 (0.0)*	2 (2.3)*
Lower extremities	2 (3.5)*	2 (15.4)*	1 (5.3)*	5 (5.7)*

Values are presented as number (%); number only; mean±standard deviation, range (median, interquartile range); or mean±standard deviation (range). Duration of disease means the period between onset and first visit to clinic. \*We rounded off the numbers to two decimal places.

lower (5.56%, n = 18) than the percentage of patients with NSV who experienced disease recurrence (23.29%, n = 73). They concluded that SV and NSV had distinguishing clinical characteristics<sup>9</sup>. However, a few studies have been conducted on the clinical course of SV with a more than one-year follow-up period<sup>9-12</sup>.

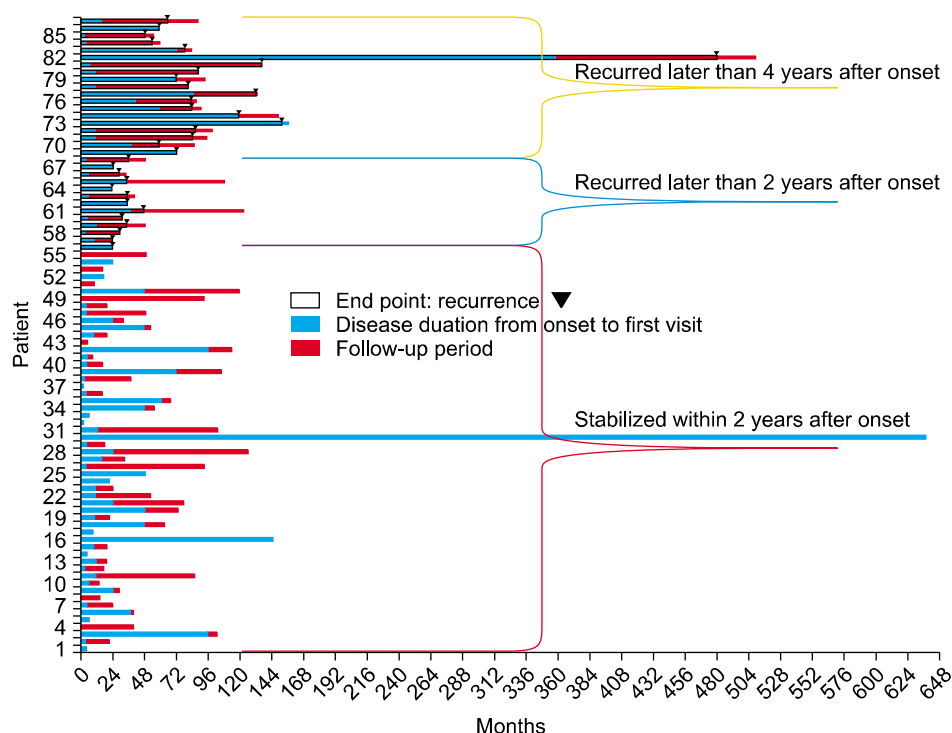
This is the first study to describe the long-term follow-up for more than a year in patients with SV, and our classifications may be used to categorize SV. Our data suggest that recurrence of SV (the appearance of new lesions or the spreading of preexisting lesions) is not uncommon even at several years after onset. On an average, our patients experienced a recurrence at 105.1 months after disease onset (Table 1, Fig. 2). We observed that 36.7% of our entire patient cohort had a recurrence of the disease after two years; and among these patients, 59.3% experienced a recurrence after four years. Among the 44 patients who were observed for more than 48 months after disease onset, 19 (43.2%) experienced a late recurrence. Although 24 patients, who were not followed up for 24 months were included in group 1, we need to closely and regularly check for the recurrence over a long time period.

Patients in the long-term dormant group 3 had more frequent and persistent involvement of the trunk or face and neck than patients in the other groups, which may be an indicator of disease prognosis. There was no significant difference in treatments used among the three groups. In

most of the cases, medical treatments (including phototherapy, topical steroids, topical tacrolimus, or short-term oral steroids) were used. When medical treatments failed, surgery was performed or the treatment was stopped.

Our study has several limitations including its retrospective design. Since we used clinical notes and photographs, we could not employ quantitative vitiligo scoring or grading systems to determine the recurrence status. The follow-up periods in our patients also varied. The patients in group 2 and group 3 were observed for a longer period than the patients in group 1 (typical rapid stabilization group), which was probably because the patients in group 1 did not seek treatment or feel the need to follow-up due to absence of disease recurrence. To overcome the limitation of this retrospective study, we think that we need to determine the minimal follow-up duration when we recruit patients with SV. As in previous studies, most of the studied patients underwent treatment for SV but were not treated with the same protocol. Therefore, the correlation between the type of treatment and the incidence of recurrence could not be established. Despite aggressive treatment, some patients experienced disease recurrence after the second year following onset. This observation supports our finding that SV may achieve stability but the disease may recur in spite of treatment years later.

This is the first study to evaluate the long-term follow-up in patients with SV. In contrast to the theory that SV tends



**Fig. 2.** Duration of the disease, length of the observation period and time to recurrence in individual cases.

to stabilize after a rapid progression within the first two years after onset, we found that many patients experienced a disease recurrence in spite of treatment. Therefore, we believe that regular assessment of SV is worthwhile, and patients should be informed that SV may recur at any time even after they achieve a stable state. Additional long-term follow-up data are needed to more accurately understand the natural course and the long-term recurrence rate of SV.

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