

Acknowledgments

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Disclosure

All authors have no conflict of interest.

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Figure legends

Fig. 1 Age distribution of disease onset in patients with RP

The mean age at onset of disease was 52.7 years old with a range from 3 to 97 years old. Older people (more than 51 years old) tend to develop RP rather than younger people (0-20 years old).

Fig. 2 Effects of immunosuppressants to the airway involvement of RP patients

Prevalence rates of airway involvement in patients with RP were 100, 42.6, 50.0 and 57.0% in treatment with steroid only, steroid with MTX, CPA, CYA and AZP, respectively, with and/or after each treatment.

Fig. 3 Summary of prognostic outcome in patients with RP in this survey

Medication was discontinued without any manifestation in 11 patients (4.6%). 159 patients (66.5%) were well controlled and, in total, 71.1% of patients in our cohort responded to the treatments. 32 patients (13.4%) showed limited response and 9 patients (3.8%) suffered from progressive disease or relapse. 22 patients (9.0%) died.

Table 1 Summary of Japan RP Questionnaire

a. Patients' profile

Sex, onset age, follow-up years and diagnostic delay

b. Clinical features

Primary and follow-up

c. Laboratory findings

Laboratory tests, image analysis and histopathologic features

d. Treatment

Non-steroidal anti-inflammatory, prednisolone, immunosuppressants, antibiotics and surgical intervention

e. Prognosis and complications

Tracheal collapse, tracheotomy, vulvar surgery and death

Table 2 Characteristics of patients with RP in McAdam series¹⁰⁾ and current survey

	McAdam (n=159)		Current survey (n=239)	
Profile				
Male-female ratio	83:76		127:112	
Mean age			57 (range 6-104)	
Mean age of disease onset	44		53 (range 3-97)	
Disease duration (yr)			5.3 (range 1-33)	
Clinical features (%)				
	Onset		Follow-up	
	Onset	Follow-up	Onset	Follow-up
External ear	26	89	57	78
Internal ear	6.4	46	3.8	27
Nasal cartilage	13	72	2.1	39
Airway	14	56	17	50
laryngo				20
tracheobronchial				41
Eye	14	65	9.2	46
conjunctivitis				15
scleritis				26
uveitis				11
Arthritis	23	81	6.2	39
Skin		17		13
Cardiovascular		24		7.1

Neurological	2.9	9.6
Renal		6.7
Myelodysplasia		1.7

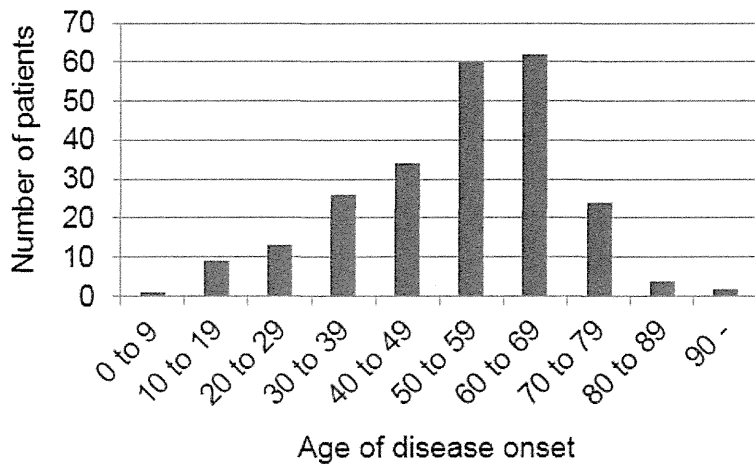


Fig. 1

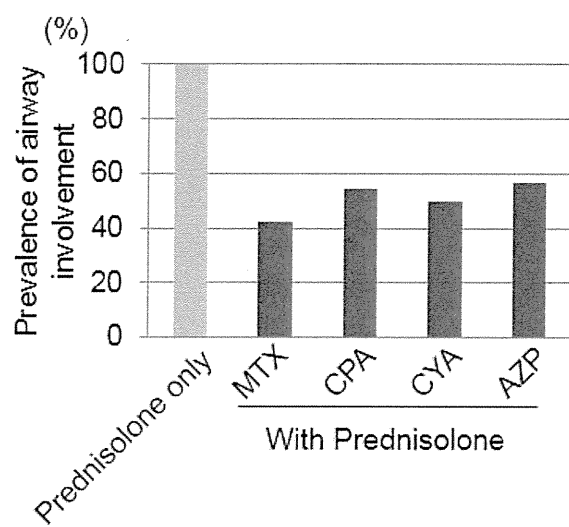


Fig. 2

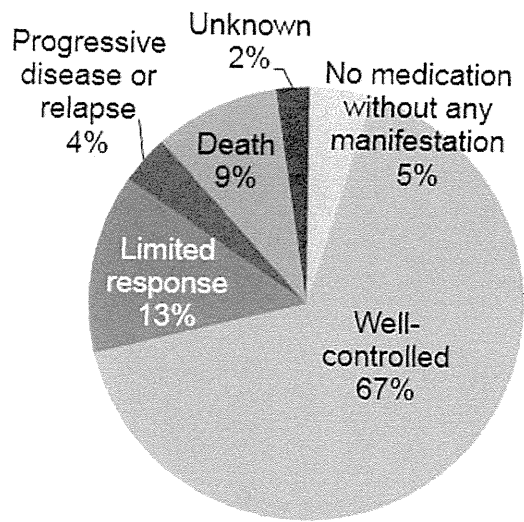


Fig. 3

V. 平成 24～25 年度班員名簿

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