

Epidemiology of Behçet's disease using the Registry of Rare Diseases of Tuscany

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INTRODUCTION

Behçet's Disease (BD) is a systemic, chronic-relapsing vasculitis of unknown etiology whose diagnosis is based on clinical criteria. The BD clinical presentation is extremely variable, and its effective management requires the prompt initiation of immunosuppressive treatments to avoid irreversible damage to vital organs

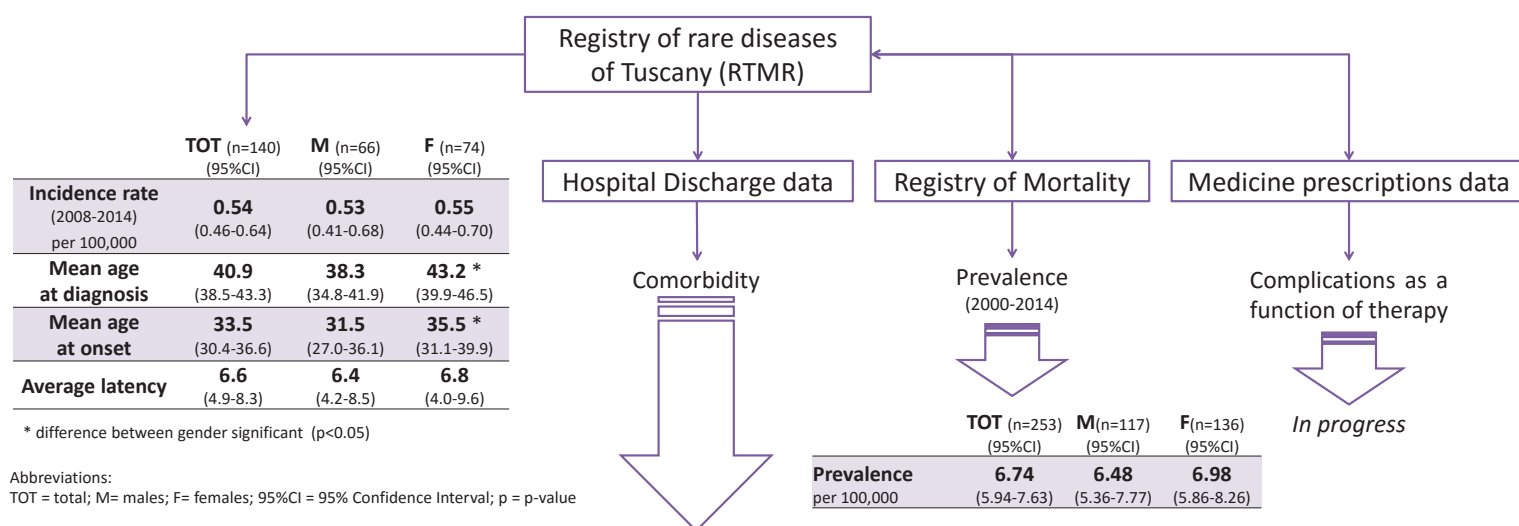
AIMS

To describe the epidemiology of BD in Tuscany (population: 3.8 million inhabitants) (Italy)

METHODS

BD cases collected by the population-based Registry of Rare Diseases were linked to registry of mortality, hospital discharge data, and medicine prescriptions data. Epidemiological indicators and causes of hospitalizations were assessed through a multi-source approach

RESULTS



Abbreviations:
TOT = total; M= males; F= females; 95%CI = 95% Confidence Interval; p = p-value

Causes of hospitalization (ICD9 CM codes)	TOT		M		F		Diff proportion test (p value)
	n	%	n	%	n	%	
Infectious And Parasitic Diseases (001-139)*	8	6.9	5	9.6	3	4.7	0.300
Neoplasms (140-239)	20	17.2	4	7.7	16	25.0	0.014
Endocrine, Nutritional And Metabolic Diseases, And Immunity Disorders (240-279)	37	31.9	11	21.2	26	40.6	0.026
Diseases Of The Blood And Blood-Forming Organs (280-289)	16	13.8	4	7.7	12	18.8	0.085
Diseases Of The Nervous System And Sense Organs (320-389)	46	39.7	17	32.7	29	45.3	0.168
Diseases Of The Nervous System (320-359)	27	23.3	6	11.5	21	32.8	0.007
Disorders Of The Eye And Adnexa (360-379)	22	19.0	12	23.1	10	15.6	0.306
Diseases Of The Circulatory System (390-459)	53	45.7	25	48.1	28	43.8	0.644
Diseases Of The Respiratory System (460-519)	27	23.3	10	19.2	17	26.6	0.348
Diseases Of The Digestive System (520-579)	46	39.7	19	36.5	27	42.2	0.533
Diseases Of The Genitourinary System (580-629)	38	32.8	14	26.9	24	37.5	0.226
Diseases Of The Skin And Subcutaneous Tissue (680-709)	20	17.2	8	15.4	12	18.8	0.630
Diseases Of The Musculoskeletal System And Connective Tissue (710-739) **	36	31.0	13	25.0	23	35.9	0.207

* excluded 1361 (Behçet's disease)

** excluded Orthopedic causes (traumatic or similar)

CONCLUSIONS

- ✓ Incidence and prevalence estimates are consistent with other European studies
- ✓ Age at diagnosis and at onset are higher in women (p<0.05)
- ✓ The study confirms the involvement of neurologic, vascular, and eye organs
- ✓ A higher percentage of hospitalizations for women was observed in all the causes except for diseases of the eye and the circulatory system

The use of a high-quality population-based rare disease registry, linkable to other health data sources, allows to provide a wide epidemiological profile of the Behçet's disease.