

MEETING ABSTRACTS

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P1

Rare Diseases in Spain: a nationwide registry-based mortality study

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Background

Rare diseases (RD) are still lacking of population-based data and epidemiological indicators. The aim of this study is to assess 15-years' time trends of mortality attributed to RD in Spain.

Methods

Mortality statistics from the Spanish National Statistics Institute provide population-based data [1]. Deaths due to RD were extracted from official annual databases (1999-2013). Only those ICD-10 codes considered as RD by SpainRDR experts were included in this study [2]. Annual sex- and age-specific adjusted mortality rates per 100,000 inhabitants were calculated and time trends were performed by joint-point regression analysis.

Results

RD mortality represents 1.2% of all registered deaths from 1999 to 2013 in Spain. Mortality attributed to RD is higher in males (51.2%) than females (48.8%). Children (<15 years old) account for 15.2% of deceases. Distribution of RD deaths according to main ICD-10 groups is displayed in Fig. 1.

Regarding time trends of RD mortality (Fig. 2), there has been a 0.95% decline in the annual age-adjusted death rates due to all RD (-0.95%, $p < 0.001$). In addition:

Decrease trends were also observed in the following subgroups: RD of the blood and blood-forming organs and certain rare disorders involving the immune mechanism (-2.06%, $p < 0.001$), RD of the circulatory system (-3.90%, $p < 0.01$), and rare congenital malformations, deformations and chromosomal abnormalities (-5.39%, $p < 0.01$).

Increase trends of annual age-adjusted death rates were detected for RD of the nervous system (1.85%, $p < 0.01$), RD of the respiratory system (2.39%, $p < 0.01$), RD of the digestive system (1.83%, $p < 0.05$) and those RD affecting the genitourinary system (9.38% $p < 0.05$).

Other RD groups have not showed any significant change in this period.

Conclusion

Official mortality statistics share criteria for analysing uniform and robust time series, which is useful for studying low-prevalence diseases. Assessed RD mortality trends are valuable information for the health authorities in Spain.

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References

1. National Statistics Institute (Instituto Nacional de Estadística), Spanish Statistical Office [<http://www.ine.es/en/>]
2. Spanish Rare Diseases Registries Research Network (SpainRDR) [<https://spainrdr.isciii.es/en/>]

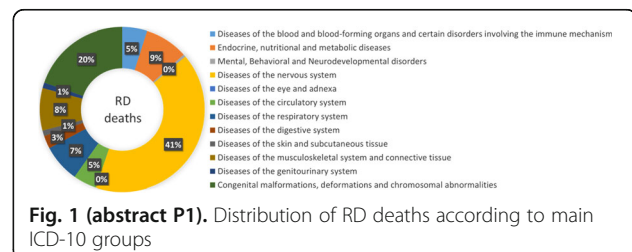


Fig. 1 (abstract P1). Distribution of RD deaths according to main ICD-10 groups

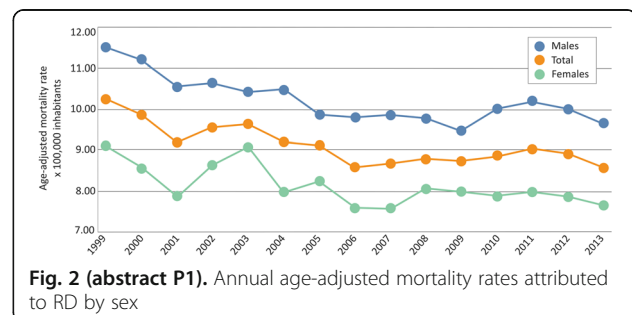


Fig. 2 (abstract P1). Annual age-adjusted mortality rates attributed to RD by sex

