Global incidence and mortality of idiopathic pulmonary fibrosis:

a systematic review

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As idiopathic pulmonary fibrosis emerges as an important public health problem, there is a

need to coordinate data on incidence and mortality globally. This study aims to systematically

assess all available studies to investigate the global burden of disease.

Medline and Embase databases were searched systematically for all population-based studies

of incidence or mortality of idiopathic pulmonary fibrosis. Clinical case series and prevalence

studies were excluded. The search was supplemented using the Google search engine, hand-

searching of references and conference abstracts. Data were extracted independently by two

authors using a pre-specified proforma, with assessment of methodological quality.

34 studies were identified, providing data from 21 countries from 1968-2012. 28 studies

reported incidence data and eight reported mortality data. In studies from the year 2000

onwards, we estimated a conservative incidence range of 3-9 cases per 100000 per year for

Europe and North America. Incidence was lower in East Asia and South America. The

majority of studies showed an increase in incidence over time.

The incidence of idiopathic pulmonary fibrosis is increasing worldwide and rates are coming

together across countries. Current data suggest incidence is similar to that of conditions such

as stomach, liver, testicular and cervical cancers.