Disease registers in England

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Abstract

The importance of accurate recording and monitoring the occurrence of disease is well recognised and there is a long tradition of the establishment of disease registers in England with over 200 in existence. These registers provide high quality information to: inform patient care, improve public health, advance medical knowledge and support clinical practice.

This article provides an overview of disease registers focussing on population-based registers of cerebral palsy, childhood cancer, congenital anomaly and diabetes. The success of these registers is dependent on a number of factors including an extensive local network of notifiers, the use of multiple source case ascertainment, adopting consistent recording and coding practices. The paediatrician can play an important role in supporting registers through the timely notification of cases and reviewing cases to ensure accurate diagnoses are recorded.

Keywords epidemiology; incidence; population-based; prevalence; registries

Introduction

What is a disease register?

A disease register is a documentation of all cases of a certain disease or health condition, which occur within a defined population. Registers are held by registries, which are the systems in place for the continuous registration of cases. In general, clinical sources prospectively notify new cases as they arise in the clinical setting. Along with personal information, clinical data that is routinely collected in clinical practice, is submitted, with the notified variables differing by registry and disease of interest. To maximise case ascertainment, data is submitted from multiple sources.

Disease registers can be hospital-, clinic- or population-based. Hospital- and clinic-based registers tend to contain data on all patients with the specific disease registered or treated in a particular hospital or clinic. These registers can be disadvantaged in that the denominator for the base population is commonly unknown, and ascertainment may be biased due to different characteristics or behaviours of cases that are and are not registered or treated at the included hospital/clinic. Population-based registers, with registration dependent on place of residence, are notified of data on cases occurring within a geographically defined area. The advantage of the population-based design is the availability of denominator data and a high ascertainment of all cases regardless of their characteristics. Despite population-based registers being more financially and resource intensive, they are considered the most valuable type of disease register.

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Purpose of disease registers

There are four clear purposes of disease registers: to improve patient care, to enhance public health, to advance medical knowledge and to better information provision. From a patient care perspective, information held by disease registers can be used to monitor high risk groups, manage demand for care, identify the correct patient care pathways and identify delays in diagnosis. In public health terms, disease registers undertake surveillance of the particular condition in the population, help inform the planning of healthcare provision for that condition, and can monitor health burden and the impact of healthcare interventions. Disease registers also contribute to advancing medical knowledge through research into the patterns and causes of disease. Disease registers can also provide information to clinicians to support their clinical practice. In her 2011 annual report, the Chief Medical Officer for England commented that: *"Surveillance and studying the epidemiology of diseases have been a driving force behind improvements to public health. They help to identify and prioritise where action is needed, characterise and inform our understanding of the drivers affecting public health and monitor the effectiveness of our interventions"*. She went on to recommend: *"Public Health England needs to develop and implement a set of coherent national surveillance systems for non-communicable diseases, congenital anomalies and important medical, environmental and lifestyle risk factors"*.

What makes a successful register?

There are a number of factors which are essential for the successful running of a disease register. It is important to have a clear case definition so that only relevant cases are included. This should, where possible, be based on agreed international definitions. Having predefined aims will ensure that the register is collecting data on the required variables. The collection of unnecessary data can produce a lack of quality for the core variables due to the time pressures of the clinical personnel who notify the registers. The data collection system should relate to the function and aims of the register. Clear leadership is needed from the outset but having a multidisciplinary team involved with the register, including a clinician with expertise in the disease of interest, expert coders, a computer scientist and a biostatistician will facilitate the successful running of the register. Using multiple sources of data increases case ascertainment and allows for cross-checking. Close working links between clinicians who are responsible for notifying cases and the register staff are vital in order to maximise case ascertainment. Where possible, the adoption of internationally approved methods of coding, recording, and analysis will contribute to the quality of the data collected. Ethical issues related to confidentiality, patient consent and data protection should be outlined at the onset of the registry and regularly reviewed and updated. One of the most significant challenges to disease registers is funding and ensuring that funding is maintained over time, and is continuous to avoid breaks in registrations, is always a challenge.

Disease registers

There are a number of disease and disease-specific registers in England covering such diverse conditions as cystic fibrosis and ankylosing spondylitis. There is likely to be considerable heterogeneity in almost every aspect of these registers in terms of purpose, size, cost, funding source and quality. However, the main principles discussed in the previous section will apply to all. In this article, examples of population-based disease registers will be detailed rather than clinical registers or databases, with particular reference to cerebral palsy, childhood cancer, congenital anomaly and childhood diabetes registers.

Cerebral palsy registers

North of England Collaborative Cerebral Palsy Survey (NECCPS): the NECCPS prospectively records data on all infants with cerebral palsy born to mothers resident in the Northern region of England (comprising the counties of North Cumbria, Northumberland, Tyne and Wear, Durham and Darlington and Teesside) from 1991. Cerebral palsy is classified according to the agreed classification system developed by the Surveillance of Cerebral Palsy in Europe, a collaborative network of cerebral palsy registers and surveys in 18 centres in 16 countries, and classed as spastic cerebral palsy (unilateral or bilateral), dyskinetic and ataxic. Cases are notified to the survey by District Convenors, who are consultant community paediatricians. They coordinate services for such children and receive information about children needing services from other paediatricians, paediatric neurologists, physiotherapists, speech therapists and the regional child development centre. The convenor completes a notification form. Further details are forwarded to the survey when the child reaches 5 years of age to confirm the diagnosis and provide details of associated impairments. Parents are invited to return a lifestyle assessment questionnaire which provides a measure of the impact of the cerebral palsy on the child and family. This questionnaire covers six dimensions: physical independence, mobility, clinical burden, schooling, economic burden and social integration, from which a global Lifestyle Assessment Score (LAS) is derived. The LAS is expressed as a percentage, with a maximally disadvantaged child scoring 100%.

It is very unusual for a case of cerebral palsy to be diagnosed after age six. However, the process of ascertainment by the convenor and the requirement to obtain parent consent means that sometimes children are added to the register up to age eight, even though diagnosed a year or two earlier. High case ascertainment is ensured because cases are notified from multiple sources, there is a regional network of interested clinicians and close links with other long-standing prospective registers (Perinatal Mortality Survey and the Northern Congenital Abnormality Survey (NorCAS)) held on the same linked database at the PHE Regional Maternity Survey Office in Newcastle. Every case of cerebral palsy mentioned on a child death certificate and every case mentioned as co-morbidity on a late notification of a congenital anomaly is ascertained by the survey.

The NECCPS is a consented register. Parents give permission for their child's information to be held on the database and reports of analysis of anonymised data to be undertaken. Parents know what data are held about their child and contribute to the NECCPS data by completing the LAS.

Information held by the NECCPS has been used in a range of studies including those investigating trends over time; rates by birth weight, gestational age and severity of cerebral palsy; and a multisite audit investigating variations in healthcare for children and young people with cerebral palsy. Disease registers can also act as a case identification system. Children notified to the NECCPS contributed to a large, European study (SPARCLE) of quality of life of children with cerebral palsy. This study demonstrated that pain is a major determinant of participation and quality of life for children with cerebral palsy and that parents and carers may under-report the presence of pain compared to the reports of the children themselves. This work, described as '*transformationally important*' by the *Lancet*, has demonstrated that by manipulating environments and involving children in decisions about their care, profound improvements to their lives can be achieved. The NECCPS is also informing the translational agenda; an ongoing programme of research, funded by the National Institute of Health Research, to identify the features of transitional care that are potentially effective and efficient for young people with complex health needs making their transition from child to adult services, is using the NECCPS to recruit young people with cerebral palsy to the study.

The NECCPS is funded by Public Health England (PHE). Until recently, there were two other cerebral palsy registers in England, one covering Merseyside and Cheshire, and one covering Oxfordshire, Berkshire, Buckinghamshire and Northamptonshire, but due to a lack of funding these registers have had to close.

Congenital anomaly registers

Local congenital anomaly registers have been established in a number of regions within the British Isles, to meet local needs and to carry out a variety of audit and research projects including audits of prenatal diagnosis and research into the causes of specific anomalies within their locality. There are currently seven regional registers in England covering the North east and North Cumbria (NorCAS), Yorkshire and Humber (YHCAR), West Midlands (WMCAR), Wessex (WANDA), East Midlands and South Yorkshire (EMSYCAR), Oxfordshire, Berkshire and Buckinghamshire (CAROBB), and the South West (SWCAR) regions (see Figure 1). These registers cover 50% of all births in England. There are also three registers in Ireland and one for all of Wales. These registers all belong to the British Isles Network of Congenital Anomaly Registers (BINOCAR) and most are also members of the European Surveillance of Congenital Anomalies (EUROCAT). Collation of the data from the regional registers is undertaken at the BINOCAR Hub located in the Wolfson Institute, Queen Mary, University of London. From April 2013, funding for the English registers has been provided by PHE.

The aim of BINOCAR is to provide continuous epidemiological monitoring of the frequency, nature and outcomes of congenital anomalies for the population of the British Isles by means of national, regional and disease-specific registers of congenital anomalies. The objectives are to undertake surveillance and analysis of congenital anomalies; monitoring and audit of health provision, detection and outcomes for congenital anomalies; provision of information to support planning and administration of the provision made for health and social care for pregnancies and infants affected by congenital anomalies;