

Disease specific registers – National Down Syndrome Cytogenetic Register (NDSCR)

- In 2009, there were 1,887 notifications of Down syndrome (27 per 10,000 total births), 163 of Patau syndrome (2 per 10,000 total births) and 506 of Edwards syndrome (7 per 10,000 total births) in England and Wales.
- There is regional variation in the prevalence of Down, Patau and Edwards syndromes with the south having a higher prevalence than the north probably reflecting the different maternal age distributions in the regions.
- Sixty-two percent of notifications of Down syndrome were diagnosed prenatally, 87% of Patau syndrome and 91% of Edwards syndrome.

Disease specific registers – Cleft lip and palate (CRANE) database

- In 2009, the CRANE database registered 878 children born with a cleft lip and/or palate in England, Wales and Northern Ireland.
- Using Hospital Episode Statistics data for England and the CRANE data for Wales and Northern Ireland the estimated incidence of cleft lip and/or palate was 15.2 per 10,000 live births (1 in 658 live births).

Funding

The Healthcare Quality Improvement Partnership (HQIP) has commissioned the collation of data in this report.

BINOCAR registers are all funded separately. CARIS (Wales) is funded by Public Health Wales, CAROBB (Thames Valley), NDSCR (National Down Syndrome Cytogenetic Register) and NorCAS (Northern England) are funded by HQIP, CRANE (cleft lip and palate database) is funded by Specialist Commissioners, EMSYCAR (East Midlands & South Yorkshire) is funded by the individual Primary Care Trusts (PCTs) and WANDA (Wessex) is funded by the Wessex Clinical Genetics Service.



British Isles Network of Congenital Anomaly Registers

Executive Summary: Congenital Anomaly Statistics 2009

England and Wales

December 2011



For more information about BINOCAR go to - www.binocar.org
or to see the full report go to - www.binocar.org/publications/reports

website: www.binocar.org

link to full report: www.binocar.org/publications/reports

Introduction

- This report collates data from five regional congenital anomaly registers, which together cover 28% of the population of England and Wales, to provide an estimate of the prevalence of congenital anomalies. More detailed information is available from the website (www.binocar.org).
- Information from the National Down Syndrome Cytogenetic Register (NDSCR) for England and Wales and the cleft lip and palate (CRANE) database for England, Wales and Northern Ireland are presented separately.
- Trends in congenital anomalies cannot be obtained by comparing earlier data from the Office for National Statistics National Congenital Anomaly System (ONS NCAS) with the 2009 data in this report; the prevalence reported by the British Isles Network of Congenital Anomaly Registers (BINOCAR) is consistently higher than that reported by NCAS due to active and multiple sources of ascertainment.

Congenital anomaly notifications

- In 2009, there were 4,181 notifications of congenital anomalies to the five BINOCAR registers (East Midlands & South Yorkshire, Northern England, Thames Valley, Wessex and Wales). The prevalence was 206 per 10,000 total births (1 in 49 births).
- The prevalence of births with congenital anomalies increased between 2005 and 2006 and then showed a 17% decrease from 248 per 10,000 total births in 2006 to 206 per 10,000 total births in 2009, probably partly due to a lag in registration in later years.

Timing of diagnosis and outcome

- Fifty-three percent of diagnoses occurred prenatally in 2009. The prevalence of prenatal diagnosis has decreased from 114 per 10,000 total births in 2005 to 109 per 10,000 total births in 2009.

- Of the pregnancies in which an anomaly was suspected prenatally, 43% resulted in a termination.
- Of those born alive, where the time of diagnosis was known, 68% were diagnosed at birth, 9% were diagnosed in the first week, 7% between the 2nd and 4th weeks and 17% after the 1st month.

Key public health indicators

- The perinatal mortality rate in the United Kingdom in 2009 was 76 per 10,000 total births of whom an estimated 11% had a congenital anomaly and 3% had congenital heart disease.
- Ninety-five percent of live births notified with congenital anomalies survived to one year of age. Live births with genital anomalies had the highest survival to one year of age (99%) and live births with respiratory anomalies had the lowest (79%).
- The overall rate of termination of pregnancy for fetal anomaly was 47 per 10,000 total births (1 in 213 births). Chromosomal anomalies accounted for 21 terminations of pregnancy per 10,000 total births (1 in 476 births).

Variation by register

- There is a significant difference in the prevalence of births with congenital anomalies in the five registers. CARIS (Wales) had the highest prevalence of 275 per 10,000 total births (1 in 36 births) and EMSYCAR (East Midlands & South Yorkshire) had the lowest prevalence of 172 per 10,000 total births (1 in 58 births).
- WANDA (Wessex) had the highest rate of termination of pregnancy (65 per 10,000 total births, 1 in 154 births) and EMSYCAR (East Midlands & South Yorkshire) had the lowest rate (38 per 10,000 total births, 1 in 263 births).
- Regional variations in prevalence and rates of termination of pregnancy for fetal anomaly may be due to both differences in ascertainment as well as differences in risk factors such as maternal age.