Report:

Congenital anomaly notifications 2007, England and Wales

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Key findings

- There were 5,077 children born in England and Wales in 2007 notified to the National Congenital Anomaly System as having one or more congenital anomalies
- In 2007, 44 per cent of all births in England and Wales were to mothers living in areas covered by a local congenital anomaly register
- The notification rate for areas of England and Wales covered by a local congenital anomaly register was 131.6 per 10,000 live and stillbirths, compared with 30.1 per 10,000 for areas not covered by a local register
- The notification rate for cardiovascular anomalies in areas covered by local registers was over ten times the rate in areas without a local register

Background

The National Congenital Anomaly System (NCAS) was set up in 1964 to monitor the occurrence of congenital anomalies in England and Wales. A congenital anomaly is a structural or functional abnormality of the human body that develops before birth. Reporting of affected live and stillbirths to the system is voluntary. The main purpose of NCAS is public health surveillance, but the system also provides the only national data on the number of children born with congenital anomalies. The system is maintained by the Office for National Statistics (ONS).

In 2007 a new classification of congenital anomalies was introduced.¹ The new classification has been developed in conjunction with the British Isles Network of Congenital Anomaly Registers (BINOCAR).² The new coding framework, which is based on the European Surveillance of Congenital Anomalies (EUROCAT) guide to coding congenital anomalies,³ incorporates tighter rules for deciding which cases should be included in ONS congenital anomaly statistics.

ONS receives data from two sources: electronically from local congenital anomaly registers for the areas covered by a register, and on paper SD56 forms directly from NHS Trusts in areas without a local register.¹ In 2007 ONS received data from eight local registers. These covered 41 per cent of births in England and all births in Wales (Box One). The

North Thames (West) Congenital Malformation Register ceased sending records to NCAS in September 2007 due to loss of funding so figures for this register are incomplete.

The local registers ascertain cases of congenital anomalies from multiple sources including SD56 forms from NHS Trusts, and information from cytogenetic laboratories, neonatal units, and other hospital and community child health services. Cases can be added to NCAS at any time after the birth.

It is recognised that there is under-reporting in NCAS, particularly in the areas not covered by local registers.^{4,5} Table 1 shows the numbers and rates of notifications from 1997 to 2007 for England and Wales, the area covered by each local register, and the rest of England. Notification rates for areas now covered by local registers all increased as the registers began participating in electronic data transfer to NCAS; for example, the rate in the South West was 93.7 per 10,000 live and stillbirths in 2002 and rose to 313.5 per 10,000 in 2003, when the South West register began submitting data to NCAS. In 1997 all data submitted to NCAS was on a SD56 form; by 2007, 79 per cent of congenital anomaly notifications were for births from areas covered by local congenital anomaly registers. However, only 44 per cent of all births in England and Wales occurred within these areas.

A 2005 study compared ascertainment of anomalies on NCAS with data on four local English congenital anomaly registers, for births that occurred during the period 1991–1999.⁴ This was before those local registers started submitting data to NCAS. The overall ascertainment by NCAS was 40 per cent; ascertainment rates by condition varied from 12 per cent for cardiac defects to 75 per cent for cleft lip. The issue of under-reporting in NCAS in areas not covered by registers in NCAS was also acknowledged in the Chief Medical Officer for England's Annual Reports for 2004 and 2005.⁶

The overall operation of NCAS will be reviewed during 2009 and ONS will be consulting on the requirements for statistics on congenital anomalies.

Notifications in 2007

There were 5,077 children born in 2007 notified to NCAS (Table 1). Of these, 3,988 (79 per cent) were born to mothers resident in an area covered by a local congenital anomaly register. The notification rate for these register areas was 131.6 notifications per 10,000 live and stillbirths compared with 30.1 per 10,000 for the parts of England not covered by a local register. A comparison

Box one

Local registers exchanging data electronically with ONS

Year register began
sending data

1998	Welsh Congenital Anomaly Register and Information Service (CARIS)
1999	East Midlands & South Yorkshire Congenital Anomaly Register
2000	North Thames (West) Congenital Malformation Register ¹
2000	Merseyside and Cheshire Congenital Anomaly Survey
2002	Wessex Antenatally Detected Anomalies Register (WANDA) ¹
2002	Congenital Anomaly Register for Oxfordshire, Berkshire & Buckinghamshire (CAROBB) (Oxfordshire only prior to 2004)
2003	Northern Congenital Abnormality Register (NorCas)
2003	South West Congenital Anomaly Register

Area coverage of each local congenital anomaly register. Areas are health authorities (1996–2001) and former strategic health authorities (2002–06) unless otherwise stated. Some of the areas in 2007 used other geographic codes to delineate the registers as noted below.

Boundaries used are those in existence at the time of the birth.

East Midlands & South Yorkshire congenital anomaly register is comprised of Derbyshire, Leicestershire, Lincolnshire, Northamptonshire (since 2003), Nottinghamshire, South Yorkshire, South Humber (1996–2001), North Lincolnshire PCT (2002 onwards), North East Lincolnshire PCT (2002 onwards). Areas excluded from East Midlands & South Yorkshire register (and England & Wales as a whole) in 2006: South Derbyshire, Lincolnshire and Nottinghamshire.

North Thames (West) is comprised of Bedfordshire, Hertfordshire, Hillingdon, Barnet, Ealing, Hammersmith and Hounslow, Kensington, Chelsea and Westminster, Brent and Harrow. This register ceased sending records to NCAS in September 2007 so the count for 2007 is an underestimate.

Merseyside is comprised of Cheshire and Mersey.

Prior to 2007, Oxford is comprised of Oxfordshire, Berkshire (2004 onwards) and Buckinghamshire (2004 onwards).

From 2007, Oxford is composed of the counties of Oxfordshire and Buckinghamshire plus the Unitary Authorities of Bracknell, West Berkshire, Reading, Slough, Windsor & Maidenhead, Wokingham and Milton Keynes.

Prior to 2007, Wessex is comprised of Hampshire, Isle of Wight, Dorset, Salisbury LA (1996–2001), South Wiltshire PCT (2002 onwards).

From 2007, Wessex is comprised of the Unitary Authorities of Poole, Isle of Wight, Bournemouth, Portsmouth and Southampton plus the counties of Dorset, Hampshire (excluding parts of Hart and East Hampshire and the whole of Rushmore) plus the district of Salisbury and parts of the districts of Arun and Chichester.

Prior to 2007, Northern is comprised of Northumberland, Tyne and Wear, County Durham, Tees Valley, North Cumbria (1995–2001), Carlisle PCT (2002 onwards), Eden Valley PCT (2002 onwards), West Cumbria PCT (2002 onwards).

From 2007, Northern is composed of the new North East Strategic Health Authority plus the districts of Allerdale, Carlisle, Copeland and Eden.

Prior to 2007, South West is comprised of Avon, Cornwall, Devon, Gloucestershire, Somerset, Wiltshire excluding – Salisbury LA (1995–2001)/ South Wiltshire PCT (2002 onwards).

From 2007, South West is composed of the new Primary Care Organisations of South Gloucestershire, Plymouth Teaching, Bath & North East Somerset, Swindon, North Somerset, Gloucestershire, Bristol, Somerset, Cornwall & Isles of Scilly, Devon plus the new Wiltshire Primary Care Organisation that contains the districts of Kennet, North Wiltshire and West Wiltshire.

¹ These registers are hospital based. Denominators use area boundaries which are not necessarily exact matches to the areas covered by the registers.

of notification rates across registers shows that Wales had the highest notification rate in 2007 (223.5 notifications per 10,000 live and stillbirths).

Due to the change in the classification of congenital anomalies, figures for the total number of notifications and overall notification rates for 2007 are not comparable with those for earlier years. However, the numbers and rates for specific anomalies will be comparable, provided the International Classification of Diseases, Tenth revision codes used are unchanged. Table 2 examines the impact of adopting the new coding framework on statistics for 2006 based on notifications on NCAS when figures for 2006 were first published in January 2008.

In addition, the incomplete data from the North Thames (West) register also has an impact on time series analysis. In 2007, there were only 316 notifications from this register in comparison to 700 in the previous year.

Tables 3 and 4 show trends in the numbers and rates of notification for areas covered, and areas not covered, by a local register. In these tables a birth is counted as within a register area if the mother was living at a postcode covered by a register at the time of the birth.

Condition-specific notification rates

There were substantial differences in condition-specific notification rates between areas now covered by local registers and those without a register (Tables 3 and 4 respectively).

The largest difference was for cardiovascular anomalies, where the rate for areas covered by a register in 2007 was over ten times that for areas without a register (31.7 notifications per 10,000 live and stillbirths compared with 2.9 per 10,000 live and stillbirths). Cardiovascular anomalies are not readily apparent at birth and so may not be notified by a doctor or midwife on a SD56 form.⁴ The local registers identify anomalies which become evident later in the child's life via alternative sources.

Cleft lip is a congenital anomaly which is highly visible at birth.⁴ For this reason notifications to NCAS could be expected to be reasonably complete even for areas not covered by a local register. In 2007 the notification rate for non-register areas was less than half that of register areas (3.1 and 7.6 notifications per 10,000 live and stillbirths respectively). The size of this difference between rates for register and

Table 1

Congenital anomalies notification rates from registers¹ participating in electronic data transfer to the National Congenital Anomaly System (NCAS), 1997–2007

	1997	1998	1999	2000	2001	2002	2003	2004	2005	2006 ²	2007 ^{2,3}
Number of babies notified											
England and Wales	5,967	6,820	7,985	8,556	7,960	7,834	8,391	7,886	8,070	7,198	5,077 ⁴
Wales	533	1,547	1,379	1,518	1,383	1,339	1,311	1,235	1,162	1,076	773
East Midlands & South Yorkshire	815	703	1,648	1,583	1,689	1,610	1,229	1,142	1,288	750	615
North Thames (West)	281	319	330	493	475	440	500	600	844	700	316⁴
Merseyside	216	214	238	649	487	370	396	382	384	364	284
Oxford	31	38	44	75	29	71	78	271	383	332	217
Wessex	228	191	223	189	220	375	352	369	396	425	362
Northern	313	296	322	287	262	319	677	661	647	647	476
South West	442	443	666	568	454	396	1,383	1,423	1,404	1,362	945
Rest of England	3,108	3,069	3,135	3,194	2,961	2,914	2,465	1,803	1,562	1,542	1,089
Percentage of all births in England and Wales covered by a local congenital anomaly register	–	5.3	14.4	26.7	25.9	32.8	41.0	44.0	44.0	43.9	43.7
Rates per 10,000 live and stillbirths											
England and Wales	92.3	106.7	127.8	140.9	133.2	130.7	134.3	122.6	124.3	111.6	76.3
Wales	153.6	460.1	427.4	482.7	449.4	440.9	415.4	379.9	354.6	318.3	223.5
East Midlands & South Yorkshire	136.2	118.1	286.9	285.0	311.0	294.9	191.1	172.1	191.3	179.0	141.7
North Thames (West)	56.4	64.0	66.6	100.4	106.5	89.2	97.9	114.6	158.6	127.0	55.4
Merseyside	77.2	77.8	89.6	250.8	194.0	149.6	153.4	144.4	144.9	133.3	102.9
Oxford	41.2	50.8	59.6	103.6	41.1	102.3	104.5	97.7	138.4	114.3	73.0
Wessex	79.2	66.4	80.4	70.6	84.6	144.3	130.6	133.2	142.6	146.0	125.0
Northern	94.6	91.9	103.5	96.4	90.2	108.5	223.2	211.8	204.7	198.1	144.0
South West	95.1	101.9	149.1	132.8	115.2	93.7	313.5	317.5	310.4	290.3	194.3
Rest of England	86.8	86.1	90.0	94.3	86.8	86.8	71.8	54.0	46.4	44.1	30.1

1 See Box One for details of areas covered by a local register.

2 Figures for 2006 and 2007 exclude data for some areas which used to be part of East Midlands & South Yorkshire congenital anomaly register.

3 A new exclusion list was implemented in 2007. Some minor anomalies excluded from counts for 2007 would have been included in figures for earlier years.

4 The North Thames (West) register ceased sending records to NCAS in September 2007 so the figures for 2007 are an underestimate.

Note: Data in bold indicate the years that the registers have been sending data to NCAS.

Source: National Congenital Anomaly System at 25 September 2008

Table 2

Impact of implementing BINOCAR Coding Framework on counts of notifications on National Congenital Anomaly System (NCAS)

Year	Date of extract	Number of babies on NCAS	Overall notification rate ¹	Cases with at least one cause in the Framework	Per cent excluded	New notification rate ¹
Areas covered by a register						
2006	17 October 2007	5,123	173.2	4,458	13.0	150.7
2007	25 September 2008	4,416	145.8	3,988	9.7	131.6
Areas not covered by a register						
2006	17 October 2007	1,519	43.5	1,161	23.6	33.2
2007	25 September 2008	1,335	36.9	1,089	18.4	30.1

1 Rate per 10,000 live and stillbirths.

Source: National Congenital Anomaly System

non-register areas for this very visible condition suggests that some cases in non-register areas are not being reported to NCAS.

Other patterns in the data

Congenital anomaly notification rates were highest among children of mothers over 40 years old.¹ In areas covered by congenital anomaly registers the notification rate in 2007 for mothers under 20 years old was 153.0 per 10,000 live and stillbirths. Mothers aged 35 to 39 had the lowest notification rate (121.0 notifications per 10,000 live and stillbirths) and those aged over 40 years the highest (172.1 per 10,000). In areas not covered by a congenital anomaly register, mothers aged over 40 years also had the highest notification rate.

Notification rates were also higher for children who were part of multiple births.¹ In areas covered by local registers the notification rate for children who were part of a multiple birth in 2007 was 172.0 per 10,000 live and stillbirths, compared with 126.3 per 10,000 for singletons.

Research⁷ linking congenital anomaly notifications to birth registrations between 1997 and 2001 showed that 14 per cent of birth registrations classified as part of a multiple birth were not recorded as such on their congenital anomaly registration. Only 2 per cent of registrations which were singleton births were not recorded as such on NCAS. It is not known whether this systematic bias in NCAS notifications still exists; if so the notification rate for multiple births is likely to be an underestimate.

Table 3

Congenital anomaly notifications, selected conditions, 2000–07¹England and Wales: Register areas²Numbers and rates³

	2000		2001		2002		2003		2004		2005		2006 ⁴		2007 ^{4,5}	
	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate
All babies notified using old exclusion list	4,243	261.9	4,034	260.6	4,205	213.7	5,926	231.2	6,083	215.0	6,508	228.0	5,656	191.2	4,416	145.8
All babies notified using new exclusion list ⁶															3,988	131.6
Babies with a mention of:																
Central nervous system anomalies	215	13.3	198	12.8	255	13.0	289	11.3	365	12.9	352	12.3	316	10.7	226	7.5
Neural tube defects	48	3.0	36	2.3	62	3.2	70	2.7	81	2.9	61	2.1	53	1.8	55	1.8
Anencephalus	10	0.6	7	0.5	17	0.9	19	0.7	18	0.6	16	0.6	10	0.3	12	0.4
Spina bifida	32	2.0	21	1.4	38	1.9	43	1.7	52	1.8	34	1.2	36	1.2	36	1.2
Hydrocephalus	57	3.5	37	2.4	47	2.4	56	2.2	67	2.4	82	2.9	87	2.9	66	2.2
Cleft lip	117	7.2	135	8.7	145	7.4	226	8.8	242	8.6	257	9.0	199	6.7	229	7.6
Cleft palate ⁶	91	5.6	91	5.9	96	4.9	120	4.7	137	4.8	165	5.8	133	4.5	115	3.8
Cardiovascular anomalies ⁶	949	58.6	823	53.2	823	41.8	1,385	54.0	1,334	47.1	1,470	51.5	1,290	43.6	961	31.7
Hypospadias	273	16.4	299	18.8	268	13.5	394	15.1	389	13.4	496	17.0	447	14.8	342	11.3
Limb reductions	77	4.8	79	5.1	82	4.2	124	4.8	160	5.7	140	4.9	128	4.3	101	3.3
Gastroschisis	51	3.1	54	3.5	55	2.8	90	3.5	152	5.4	105	3.7	134	4.5	94	3.1
Down syndrome	166	10.2	165	10.7	165	8.4	276	10.8	366	12.9	379	13.3	319	10.8	243	8.0

1 Figures for 1998 and 1999 are omitted because they would be based on data from only two registers.

2 See Box One for details of areas covered by a local register.

3 Notification rate per 10,000 live and stillbirths.

4 Figures for 2006 and 2007 exclude data for some areas which used to be part of East Midlands & South Yorkshire congenital anomaly register.

5 The North Thames (West) register ceased sending records to NCAS in September 2007 so the figures for 2007 are an underestimate.

6 A new exclusion list was implemented in 2007. Some minor anomalies excluded from counts for 2007 would have been included in figures for earlier years.

Note: Rates calculated from fewer than 20 anomalies are distinguished by italic type as a warning to the user that their reliability as a measure may be affected by the small number of events.

Source: National Congenital Anomaly System at 25 September 2008

Table 4

Congenital anomaly notifications, selected conditions, 1997, 2000–07¹England and Wales: Non-register areas²Numbers and rates³

	1997		2000		2001		2002		2003		2004		2005		2006		2007	
	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate
All babies notified using old exclusion list	5,967	92.3	4,313	96.9	3,926	88.7	3,629	90.2	2,465	66.9	1,803	50.1	1,562	42.9	1,542	44.1	1,335	36.9
All babies notified using new exclusion list ⁴																	1,089	30.1
Babies with a mention of:																		
Central nervous system anomalies	220	3.4	199	4.5	188	4.2	176	4.4	131	3.6	86	2.4	98	2.7	66	1.9	58	1.6
Neural tube defects	80	1.2	75	1.7	65	1.5	53	1.3	54	1.5	38	1.1	37	1.0	23	0.7	20	0.6
Anencephalus	27	0.4	17	0.4	20	0.5	13	0.3	14	0.4	9	0.2	9	0.2	6	0.2	9	0.2
Spina bifida	48	0.7	48	1.1	39	0.9	34	0.8	41	1.1	25	0.7	24	0.7	14	0.4	9	0.2
Hydrocephalus	64	1.0	49	1.1	50	1.1	32	0.8	33	0.9	17	0.5	22	0.6	14	0.4	20	0.6
Cleft lip	379	5.9	277	6.2	237	5.4	217	5.4	146	4.0	120	3.3	96	2.6	115	3.3	111	3.1
Cleft palate ⁴	202	3.1	127	2.9	111	2.5	110	2.7	81	2.2	54	1.5	55	1.5	43	1.2	33	0.9
Cardiovascular anomalies ⁴	666	10.3	474	10.6	454	10.3	462	11.5	195	5.3	162	4.5	148	4.1	168	4.8	105	2.9
Hypospadias	471	7.3	368	8.3	340	7.7	288	7.1	194	5.2	148	4.1	111	3.0	113	3.1	90	2.5
Limb reductions	137	2.1	130	2.9	113	2.6	100	2.5	78	2.1	61	1.7	44	1.2	53	1.5	44	1.2
Gastroschisis	89	1.4	67	1.5	59	1.3	61	1.5	40	1.1	46	1.3	27	0.7	25	0.7	24	0.7
Down syndrome	411	6.4	244	5.5	215	4.9	192	4.8	132	3.6	112	3.1	109	3.0	110	3.1	96	2.7

1 Figures for 1998 and 1999 are omitted because the corresponding figures for register areas are based on data from only two registers.

2 See Box One for details of areas covered by a local register.

3 Notification rate per 10,000 live and still births.

4 A new exclusion list was implemented in 2007. Some minor anomalies excluded from counts for 2007 would have been included in figures for earlier years.

Note: Rates calculated from fewer than 20 anomalies are distinguished by italic type as a warning to the user that their reliability as a measure may be affected by the small number of events.

Source: National Congenital Anomaly System at 25 September 2008

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