

Statistical Bulletin

Congenital anomaly notifications



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Coverage: England and Wales Theme: Health and Care

There were 4,254 children born in England and Wales in 2008 notified to the National Congenital Anomaly System as having one or more congenital anomalies. In 2008, 37 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 133.7 per 10,000 live and stillbirths, compared with 20.5 per 10,000 for areas not covered by a local register. The notification rates for cardiovascular anomalies and Down syndrome in areas covered by local registers was over fourteen times the rates 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 Eurocat guide to coding congenital anomalies³, 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. In 2008 ONS received data from seven local registers (Box One). These covered 32 per cent of births in England and all births in Wales.



BOX 1 Local registers exchanging data electronically with ONS

Year register began	Register
sending data	
1998	Welsh Congenital Anomaly Register and Information Service (CARIS)
1999	East Midlands and 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 (2002-2006) 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 the NCAS in September 2007 so the count for 2007 is an underestimate, and there are no data for 2008.

Merseyside is comprised of Cheshire and Mersey.

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



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 1998 to 2008 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 315.1 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 2008, 79 per cent of congenital anomaly notifications were for births from areas covered by local congenital anomaly registers. However, only 37 per cent of all births in England and Wales occurred within these areas.

Table 1 Congenital anomalies notification rates from registers participating in electronic data transfer (1998-2008)	
Number of babies notified to the National Congenital Anomaly System (NCAS)	

number of babies nouned to the national Congernial Anomaly system (IICAS)													
	<u>1998</u>	<u>1999</u>	2000	2001	2002	2003	2004	2005	2006 ²	2007 2,3	2008 ^{2,3}		
England and Wales	6,857	7,990	8,562	8,029	7,842	8,421	7,964	8,183	7,460	5,931	4,254		
Wales	1,578	1,379	1,518	1,438	1,339	1,321	1,274	1,215	1,210	898	837		
East Midlands & South Yorkshire	703	1,648	1,583	1,689	1,610	1,229	1,142	1,289	753	783	563		
North Thames (West) +5	319	330	493	478	440	500	600	844	700	325	-		
Merseyside	215	238	649	489	370	396	382	383	364	308	201		
Oxford	38	44	75	30	71	78	274	389	347	328	203		
Wessex	193	223	189	220	376	355	376	401	453	399	366		
Northern	297	326	293	268	325	684	669	665	674	569	514		
South West	443	667	568	456	396	1,390	1,444	1,433	1,411	1,183	693		
Rest of England	3,071	3,135	3,194	2,961	2,915	2,468	1,803	1,564	1,548	1,138	877		
Percentage of all births in England and Wales covered by a local congenital anomaly register ⁶	5.3	14.4	26.7	25.9	32.8	41.0	44.0	44.0	45.9	45.6	37.1		
Rates per 10,000 live and stillbirths													
England and Wales	107.3	127.9	141.0	134.4	130.9	134.8	123.8	126.1	115.6	89.2	62.4		
Wales	469.4	427.4	482.7	467.3	440.9	418.5	391.9	370.8	358.0	259.7	234.0		
East Midlands & South Yorkshire	118.1	286.9	285.0	311.0	294.9	191.1	172.1	191.5	179.8	180.4	127.7		
North Thames (West) *5	64.0	66.6	100.4	107.1	89.2	97.9	114.6	158.6	127.0	57.0	-		
Merseyside	78.1	89.6	250.8	194.8	149.6	153.4	144.4	144.5	133.3	111.6	71.2		
Oxford	50.8	59.6	103.6	42.5	102.3	104.5	98.8	140.5	119.4	110.4	66.2		
Wessex	67.1	80.4	70.6	84.6	144.7	131.8	135.7	144.4	155.7	137.8	122.0		
Northern	92.2	104.8	98.4	92.2	110.6	225.5	214.3	210.4	206.4	172.1	152.0		
South West	101.9	149.3	132.8	115.7	93.7	315.1	322.2	316.9	300.8	243.2	138.5		
Rest of England	86.1	90.0	94.3	86.8	86.8	71.9	54.0	46.4	44.3	31.5	20.5		

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

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

Source: National Congenital Anomaly System at 20 October 2009

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

² Figures for 2006, 2007 and 2008 exclude data for some areas which used to be part of the East Midlands and South Yorkshire congenital anomalies register.

³ A new exclusion list was implemented in 2007. Some minor anomalies excluded from counts in 2007 and 2008 would have been included in figures from earlier years.

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

⁵ All data for the North Thames (West) register in 2008 has been classified as non register and added to the Rest of England data

⁶ Due to a processing error the previously published figures for 2006 and 2007 were incorrect. These have now been corrected



The overall operation of NCAS was reviewed during 2009/2010 and the ONS are consulting on the requirements for statistics on congenital anomalies from January to March 2010.7

Notifications in 2008

There were 4,254 children born in 2008 notified to NCAS (Table 1). Of these, 3,377 (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 133.7 notifications per 10,000 live and stillbirths compared with 20.5 per 10,000 for the parts of England not covered by a local register. A comparison of notification rates across registers shows that Wales had the highest notification rate in 2008 (234.0 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 onwards are not comparable with those for earlier years. However, the numbers and rates for specific anomalies will be comparable, provided the ICD10 codes used are unchanged.

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. In 2008 the North Thames (West) register was classified as a non register and in Table 1 will show under 'rest of England'.

Tables 2 and 3 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 2 and 3 respectively).

The largest difference was for cardiovascular anomalies, where the rate for areas covered by a register in 2008 was nearly 15 times that for areas without a register (36.9 notifications per 10,000 live and stillbirths compared with 2.5 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 2008 the notification rate for non-register areas was less than a third of that for register areas (2.0 and 6.7 notifications per 10,000 live and stillbirths respectively). The size of this difference between rates for register and non-register areas for this very visible condition suggests that some cases in non-register areas are not being reported to NCAS.



Table 2	Congenital anomaly notifications, selected conditions, 2000-2008 ¹																	
England and Wales: Register areas ²																lumbe	ers and r	rates³
	200	0	2001		2002		200	3	2004		2005		2006 *		2007 +,5,5		2008	4,5,5
	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 All babies notified using new exclusion list	4,243	261.9	4,175	269.7	4,206	213.8	5,953	232.3	6,161	217.7	6,619	231.9	5,912	199.8	5,260 4,793	173.6 158	3,377	134
Babies with a mention of:																		
Central nervous system anomalies	197	12.2	184	11.9	227	11.5	270	10.5	351	12.4	335	11.7	324	11.0	272	9.0	175	6.9
Neural tube defects	48	3.0	36	2.3	62	3.2	70	2.7	81	2.9	61	2.1	55	1.9	56	1.8	47	1.9
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	13	0.5
Spina bifida	32	2.0	21	1.4	38	1.9	43	1.7	52	1.8	34	1.2	38	1.3	37	1.2	32	1.3
Hydrocephalus	57	3.5	37	2.4	47	2.4	54	2.1	68	2.4	83	2.9	92	3.1	82	2.7	59	2.3
Cleft lip	117	7.2	136	8.8	145	7.4	226	8.8	242	8.6	259	9.1	199	6.7	253	8.4	169	6.7
Cleft palate	85	5.2	95	6.1	92	4.7	115	4.5	131	4.6	160	5.6	132	4.5	127	4.2	113	4.5
Cardiovascular anomalies	908	56.1	774	50.0	785	39.9	1,340	52.3	1,290	45.6	1,464	51.3	1,370	46.3	1,260	41.6	932	36.9
Hypospadias	265	16.4	294	19.0	267	13.6	389	15.2	400	14.1	488	17.1	478	16.2	388	12.8	258	10.2
Limb reductions	77	4.8	81	5.2	82	4.2	126	4.9	162	5.7	140	4.9	126	4.3	108	3.6	63	2.5
Gastroschisis	51	3.1	55	3.6	55	2.8	90	3.5	152	5.4	105	3.7	134	4.5	97	3.2	111	4.4
Down syndrome	166	10.2	168	10.9	165	8.4	276	10.8	367	13.0	380	13.3	320	10.8	265	8.7	225	8.9

- 1 Figures for 1998 and 1999 are omitted because they would be based on data from only 2 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 Figures for 2006, 2007 and 2008 exclude data for some areas which used to be part of the East Midlands and South Yorkshire congenital anomalies register
- 5 The North Thames (West) register ceased sending records to NCAS in September 2007 so the figures for 2007 are an underestimate
- All data for the North Thames (West) register in 2008 has been classified as non register and added to the Rest of England data
- 6 A new exclusion list was implemented in 2007. Some minor anomalies excluded from counts in 2007 and 2008 would have been included in figures from earlier years,

Note: Rates calculated from fewer than 20 anomalies are distinguished by italic type as a warning to users that their reliability as a measure may be affected by the small number of events. Note: Numbers and rates for specific anomalies are comparable, provided the ICD10 codes used are unchanged.

Source: National Congenital Anomaly System at 20 October 2009

Congenital anomaly notifications, selected conditions, 1997,2000-2008

England and Wales: Non register areas 2

England and Wales: Hon register areas ² Humbers and rates ³																				
	1997 2000		2001		2002		2003		2004		2005		2006 *		2007 +,5,5		2008	,5,6		
	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate
All babies notified using old exclusion All babies notified using new exclusio		91.9	4,319	97.0	3,854	87.1	3,636	90.3	2,468	67.0	1,803	50.1	1,564	43.0	1,548	44.3	1,364 1,138	37.7 31.5	877	20.5
Babies with a mention of:																				
Central nervous system anomalies	204	3.2	189	4.2	177	4.0	175	4.3	131	3.6	85	2.4	97	2.7	66	1.9	60	1.7	48	1.1
Neural tube defects	80	1.2	75	1.7	64	1.4	53	1.3	54	1.5	38	1.1	37	1.0	23	0.7	22	0.6	16	0.4
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	4	0.1
Spina bifida	48	0.7	48	1.1	38	0.9	34	0.8	41	1.1	25	0.7	24	0.7	14	0.4	10	0.3	12	0.3
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	24	0.6
Cleft lip	379	5.9	277	6.2	236	5.3	217	5.4	146	4.0	120	3.3	96	2.6	115	3.3	113	3.1	86	2.0
Cleft palate	194	3.0	121	2.7	102	2.3	106	2.6	78	2.1	53	1.5	53	1.5	41	1.2	34	0.9	41	1.0
Cardiovascular anomalies	612	9.5	441	9.9	432	9.8	429	10.7	191	5.2	157	4.4	143	3.9	165	4.7	112	3.1	106	2.5
Hypospadias	472	7.3	368	8.3	342	7.7	284	7.1	192	5.2	146	4.1	110	3.0	110	3.1	97	2.7	72	1.7
Limb reductions	137	2.1	130	2.9	111	2.5	100	2.5	78	2.1	61	1.7	44	1.2	54	1.5	44	1.2	34	0.8
Gastroschisis	89	1.4	67	1.5	58	1.3	61	1.5	40	1.1	46	1.3	27	0.7	25	0.7	24	0.7	15	0.3
Down syndrome	411	6.4	244	5.5	211	4.8	192	4.8	132	3.6	112	3.1	109	3.0	110	3.1	100	2.8	65	1.5

- 1 Figures for 1998 and 1999 are omitted because they would be based on data from only 2 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 Figures for 2006, 2007 and 2008 exclude data for some areas which used to be part of the East Midlands and South Yorkshire congenital anomalies register.
- 5 The North Thames (West) register ceased sending records to NCAS in September 2007 so the figures for 2007 are an underestimate.
- All data for the North Thames (West) register in 2008 has been classified as non register and added to the Rest of England data
- 6 A new exclusion list was implemented in 2007. Some minor anomalies excluded from counts in 2007 and 2008 would have been included in figures from earlier years.
- Note: Rates calculated from fewer than 20 anomalies are distinguished by italic type as a warning to users that their reliability as a measure may be affected by the small number of events

Note: Numbers and rates for specific anomalies are comparable, provided the ICD10 codes used are unchanged.

Source: National Congenital Anomaly System at 20 October 2009

Other patterns in the data from local congenital anomaly registers only

Congenital anomaly notification rates were highest among children of mothers over 40 years old. In areas covered by congenital anomaly registers the notification rate in 2008 for mothers under 20 years old was 139.8 per 10,000 live and stillbirths. Mothers aged 30 to 34 had the lowest notification rate (116.5 notifications per 10,000 live and stillbirths) and



those aged over 40 years the highest (196.9 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 2008 was 193.3 per 10,000 live and stillbirths, compared with 127.8 per 10,000 for singletons.

Research⁸ 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.

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Background Notes

- 1. The statistics in this publication relate to the National Congenital Anomaly System (NCAS) as at 20 October 2009.
- 2. Details of the policy governing the release of new data are available from the media office.
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