



East Midlands & South Yorkshire

**Congenital Anomalies Register**

One of The Infant Mortality & Morbidity Studies

# **Congenital Anomalies in Births 2006 to 2010**

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## Background information

**Time period covered:** This report summarises congenital anomalies reported in pregnancies which ended between January 1<sup>st</sup> 2006 and December 31<sup>st</sup> 2010 inclusive in the East Midlands and South Yorkshire (EMSYCAR) region. In addition, Tables 1 – 4 provide data from 2000 – 2005 for comparison.

During this period, over 22,000 separate notifications were made to EMSYCAR and 12,810 cases (babies/foetuses) were registered. **We report here on the 8,170 of those that had at least one serious anomaly present at delivery. The cohort included 687 different anomalies, and a total of 12,684 in all.**

**EMSYCAR Geography:** In 2006, the geography of the area covered by EMSYCAR changed substantially for the second time. The 39 Primary Care Trusts, (PCTs) which had formed three separate Strategic Health Authorities (SHAs) and part of a fourth, and had constituted the EMSYCAR area since 2002, were re-organised. Six of the original 39 remained unchanged, the rest being reformed into nine larger PCTs. The data presented here follows the boundaries of the fourteen 'new' PCTs and one CTP (Care Trust Plus) which postdate October 2006. Data for the early part of 2006 has been allocated to the relevant 'new' PCT.

**Surveillance:** The National Congenital Anomaly System (NCAS) discontinued all routine anomaly surveillance from the end of 2008. EMSYCAR remains as the only resource available to perform this essential public health function for its resident population. The Register uses computer software provided through its membership of the European Surveillance Network (EUROCAT) to monitor anomalies, and every effort is made to submit current data into the EUROCAT system as it becomes available (i.e. one year in advance of the required schedule), in order for surveillance to be as timely as possible.

- The UK Registers responded to the closure of NCAS by establishing, in co-operation with EUROCAT and with initial funding from HQIP, a data hub for England and Wales, which comprises aggregated data from the English and Welsh Regional Registers (See EMSYCAR Report for 2005 – 2009). Annual Reports for the years 2011 and 2012, incorporating data from EMSYCAR, can be found on the BINOCAR website, [www.binocar.org](http://www.binocar.org).

**Anomaly definitions:** In order to make comparisons between the various datasets easier, this Report adopts, where possible, the updated definitions of anomalies and anomaly groups, used by EUROCAT and BINOCAR. The exceptions involve single anomalies (or groups) where absolute numbers within the EMSYCAR Region are too small and could permit the possible identification of individual cases. The revised groups, and the codes used to identify them, will be particularly apparent in Table 5.

**'Minor' anomalies:** The BINOCAR Registers have continued to refine the list of 'minor' anomalies for exclusion, to bring it into closer alignment with the EUROCAT list. The same working group has also addressed the issue of coding variability, both between different regional Registers, and between regional Registers and NCAS. All Registers have now adopted an agreed BINOCAR coding framework, which NCAS also used for births between January 1<sup>st</sup> 2007 and December 31<sup>st</sup> 2008. Some variation in reported anomaly rates in certain subgroups (particularly musculoskeletal and endocrine & metabolic disorders) is therefore to be expected in data from 2007 onwards.

**Table Format:** In preparing the tables presented here, anomaly rates (expressed as a rate per 10,000 births) have been preferred to exact numbers. Individual PCTs are, of course, welcome to contact EMSYCAR to request greater detail for their own area. This will be provided wherever possible, although it should be noted that, according to national guidelines, the number of anomaly cases in any given cell should always exceed a minimum of five. This is to ensure that data confidentiality is not compromised and that there is no possibility of individual cases being identified. For small areas and/or rare anomalies, this criterion may frequently not be met.

## EMSYCAR Data Summary

The **total number of births** occurring in the EMSYCAR region **has shown a steady increase** since 2001. Annual births are now in excess of 75,000.

In any given year, **over 2% of all births in the EMSYCAR region have serious, reportable anomalies**. Slightly more males (2.2%) than females (1.6%) and more multiple (3.5%) than singleton (2.1%) births are affected. These well-recognised trends are again confirmed by the data presented here.

Despite reduced funding during the period, **the number of cases reported annually to the European Surveillance System, EUROCAT, has increased steadily, and the total number of cases submitted for births between 1998 and 2010 now exceeds 17,700**. This reflects a huge amount of work, for both the EMSYCAR team and all the notifiers across the region, tracking cases and obtaining outcome data.

The previous EMSYCAR Report showed a slight fall in the absolute number of cases reported to EMSYCAR, particularly in the postnatal period, but no evidence that this was a real decrease in occurrence. Considerable efforts have therefore been made throughout the past year to encourage notifications from surgical units, neonatal units, the regional paediatric cardiac centre and paediatricians in general throughout the region, together with attempts to find alternative methods of notification to replace those lost as a result of reorganisation within individual PCTs.

Particular attention has been paid to securing the best possible reporting of postnatally detected cardiac anomalies. Although EMSYCAR rates of the 'serious' cardiac anomaly subgroup (defined as those requiring intervention within the first year of a child's life) are comparable with those recorded by other Registers, rates of the less serious cardiac conditions, and particularly those more likely to be diagnosed or confirmed after the neonatal period, are lower than elsewhere. Work is in progress to ensure that all relevant cardiac cases are reported to the Register at the appropriate time.

We are therefore cautiously optimistic that the data from 2011 onwards will demonstrate an increase in the reporting of such cases. Similar improvements will be extended to other anomaly groups.

While anomaly rates have historically varied between PCTs, Table 5 demonstrates that the **rates for individual anomalies have largely remained stable for EMSYCAR as a whole**. Comparison with rates reported elsewhere in Europe can easily be made from the EUROCAT website ([www.eurocat-network.eu](http://www.eurocat-network.eu)).

**Table 1. Number and proportion of births with one (or more) confirmed congenital anomaly, by year of birth 2000-2010**

| Year             | Total Births<br>n | Births with one or more confirmed anomaly |              | Births with multiple confirmed anomalies |              | Cases coded with anomalies of multiple aetiology |                                    |
|------------------|-------------------|---|--------------|--|--------------|--|------------------------------------|
|                  |                   | n   | Proportion % | n  | Proportion % | n  | Proportion % (births with anomaly) |
| <b>2000-2005</b> | <b>362,415</b>    | <b>8,945</b>                              | <b>2.5</b>   | <b>2,194</b>                             | <b>0.6</b>   | <b>N/A</b>                                       | <b>N/A</b>                         |
| <b>2006</b>      | <b>70,153</b>     | <b>1,756</b>                              | <b>2.5</b>   | <b>423</b>                               | <b>0.6</b>   | <b>N/A</b>                                       | <b>n/a</b>                         |
| <b>2007</b>      | <b>72,549</b>     | <b>1,724</b>                              | <b>2.4</b>   | <b>447</b>                               | <b>0.6</b>   | <b>160</b>                                       | <b>9.3</b>                         |
| <b>2008</b>      | <b>74,469</b>     | <b>1,660</b>                              | <b>2.2</b>   | <b>461</b>                               | <b>0.6</b>   | <b>138</b>                                       | <b>8.3</b>                         |
| <b>2009</b>      | <b>74,101</b>     | <b>1,622</b>                              | <b>2.2</b>   | <b>486</b>                               | <b>0.7</b>   | <b>150</b>                                       | <b>9.2</b>                         |
| <b>2010</b>      | <b>75,698</b>     | <b>1,408</b>                              | <b>1.9</b>   | <b>463</b>                               | <b>0.6</b>   | <b>123</b>                                       | <b>8.7</b>                         |
| <b>2006-2010</b> | <b>366,970</b>    | <b>8,170</b>                              | <b>2.2</b>   | <b>2,280</b>                             | <b>0.6</b>   | <b>571</b>                                       | <b>7.0</b>                         |

**Table 2. Number and proportion of births with one (or more) confirmed anomaly, by plurality and year of birth 2000-2010**

| Year             | Total Births<br>n | Singletons<br>n | Multiples<br>n | Singleton births - one or more anomaly |              | Multiple births - one or more anomaly |              | Births of unknown plurality with one or more anomaly |                              | Births of unknown plurality Proportion (% EMSYCAR cases) |  |
|------------------|-------------------|-----------------|----------------|--|--------------|---------------------------------------|--------------|--|------------------------------|--|--|
|                  |                   |                 |                | n                                      | Proportion % | n                                     | Proportion % | n  | Proportion of Total births % | Proportion   |  |
| <b>2000-2005</b> | <b>362,415</b>    | <b>382,004</b>  | <b>10,411</b>  | <b>8,391</b>                           | <b>2.2</b>   | <b>289</b>                            | <b>2.8</b>   | <b>265</b>   | <b>0.07</b>                  | <b>3.0</b>   |  |
| <b>2006</b>      | <b>70,153</b>     | <b>68,018</b>   | <b>2,135</b>   | <b>1,666</b>                           | <b>2.4</b>   | <b>56</b>                             | <b>2.6</b>   | <b>34</b>  | <b>0.05</b>                  | <b>1.9</b>   |  |
| <b>2007</b>      | <b>72,549</b>     | <b>70,507</b>   | <b>2,042</b>   | <b>1,609</b>                           | <b>2.3</b>   | <b>80</b>                             | <b>3.9</b>   | <b>35</b>  | <b>0.05</b>                  | <b>2.0</b>   |  |
| <b>2008</b>      | <b>74,469</b>     | <b>72,284</b>   | <b>2,185</b>   | <b>1,489</b>                           | <b>2.1</b>   | <b>74</b>                             | <b>3.4</b>   | <b>97</b>  | <b>0.13</b>                  | <b>5.8</b>   |  |
| <b>2009</b>      | <b>74,101</b>     | <b>71,713</b>   | <b>2,388</b>   | <b>1,434</b>                           | <b>2.0</b>   | <b>83</b>                             | <b>3.5</b>   | <b>105</b>   | <b>0.14</b>                  | <b>6.5</b>   |  |
| <b>2010</b>      | <b>75,698</b>     | <b>73,648</b>   | <b>2,050</b>   | <b>1,226</b>                           | <b>1.7</b>   | <b>52</b>                             | <b>2.5</b>   | <b>130</b>   | <b>0.17</b>                  | <b>9.2</b>   |  |
| <b>2006-2010</b> | <b>366,970</b>    | <b>357,196</b>  | <b>9,774</b>   | <b>7,424</b>                           | <b>2.1</b>   | <b>345</b>                            | <b>3.5</b>   | <b>401</b>   | <b>0.11</b>                  | <b>4.9</b>   |  |

**Table 3. Number and proportion of births with one (or more) confirmed anomaly, by infant sex and year of birth 2000 - 2010**

| Year             | Total Births<br>n | Male<br>n      | Females<br>n   | Males<br>with one or<br>more anomaly | Proportion | Females<br>with one or<br>more anomaly | Proportion | Unknown sex<br>with one or<br>more anomaly | Proportion<br>of total births (EMSYCAR cases) |
|------------------|-------------------|----------------|----------------|--------------------------------------|------------|--|------------|--|---|
|                  |                   |                |                | n                                    | %          | n                                      | %          | n  | %   |
| <b>2000-2005</b> | 362,415           | 186,049        | 176,366        | 4,903                                | 2.6        | 3,550                                  | 2.0        | 492  | 0.1 (5.5)                                     |
| <b>2006</b>      | 70,153            | 35,848         | 34,305         | 952                                  | 2.7        | 679                                    | 2.0        | 100  | 0.1 (5.7)                                     |
| <b>2007</b>      | 72,549            | 37,273         | 35,276         | 907                                  | 2.4        | 571                                    | 1.6        | 149  | 0.2 (8.6)                                     |
| <b>2008</b>      | 74,469            | 38,211         | 36,258         | 803                                  | 2.1        | 558                                    | 1.5        | 140  | 0.2 (8.4)                                     |
| <b>2009</b>      | 74,101            | 37,973         | 36,128         | 718                                  | 1.9        | 522                                    | 1.4        | 131  | 0.2 (8.1)                                     |
| <b>2010</b>      | 75,698            | 38,646         | 37,052         | 712                                  | 1.8        | 562                                    | 1.5        | 134  | 0.2 (9.5)                                     |
| <b>2006-2010</b> | <b>366,970</b>    | <b>187,951</b> | <b>179,019</b> | <b>4,092</b>                         | <b>2.2</b> | <b>2,892</b>                           | <b>1.6</b> | <b>654</b>                                 | <b>0.2 (8.0)</b>                              |

**Table 4. Birth status of EMSYCAR cases with one (or more) confirmed anomaly, by year of delivery 2000 - 2010**

|                  | Cases with<br>confirmed/<br>probable<br>anomalies<br>n | TOP         |             | Fetal Loss<br><=23+6 gest wks. |            | Stillbirth<br>>=24+0 gest wks. |            | Alive       |             | Died       |            |
|------------------|--|-------------|-------------|--------------------------------|------------|--------------------------------|------------|-------------|-------------|------------|------------|
|                  |  | n           | %           | n                              | %          | n                              | %          | n           | %           | n          | %          |
| <b>2000-2005</b> | <b>8,945</b>   | <b>1603</b> | <b>17.9</b> | <b>234</b>                     | <b>2.6</b> | <b>222</b>                     | <b>2.5</b> | <b>6361</b> | <b>71.1</b> | <b>525</b> | <b>5.9</b> |
| <b>2006</b>      | 1,756  | 329         | 18.7        | 29                             | 1.7        | 42                             | 2.4        | 1257        | 71.6        | 99         | 5.6        |
| <b>2007</b>      | 1,724  | 302         | 17.5        | 27                             | 1.6        | 25                             | 1.5        | 1278        | 74.1        | 92         | 5.3        |
| <b>2008</b>      | 1,660  | 311         | 18.7        | 41                             | 2.5        | 27                             | 1.6        | 1224        | 73.7        | 57         | 3.4        |
| <b>2009</b>      | 1,622  | 303         | 18.7        | 43                             | 2.7        | 40                             | 2.5        | 1171        | 72.2        | 65         | 4.0        |
| <b>2010</b>      | 1,408  | 299         | 21.2        | 34                             | 2.4        | 31                             | 2.2        | 980         | 69.6        | 64         | 4.5        |
| <b>2006-2010</b> | <b>8,170</b>   | <b>1544</b> | <b>18.9</b> | <b>174</b>                     | <b>2.1</b> | <b>165</b>                     | <b>2.0</b> | <b>5910</b> | <b>72.3</b> | <b>377</b> | <b>4.6</b> |

**Table 5. Number and Rates of Congenital Anomalies per 10,000 births, by year of birth 2006 - 2010**

|   |  | 2006             | 2007             | 2008             | 2009             | 2010              | TOTAL                          |
|---|--|------------------|------------------|------------------|------------------|-------------------|--------------------------------|
|   |  | 70153            | 72549            | 74469            | 74101            | 75698             | 366970                         |
|   |  | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000  | n<br>Rate/10,000<br>[95% C.I.] |
|   |  | <b>ICD-10</b>    |                  |                  |                  |                   |                                |
| <b>CENTRAL NERVOUS SYSTEM</b>                             | Q000-Q079  | 162              | 188              | 161              | 176              | 174               | <b>861</b>                     |
|   |  | 23.1             | 25.9             | 21.6             | 23.8             | 23.0              | 23.5 [21.9, 25.1]              |
|   | <b>All Neural Tube Defects</b>                         |                  |                  |                  |                  |                   | <b>433</b>                     |
|   | Q000-Q019<br>& Q050-Q059                               | 84               | 86               | 78               | 82               | 103               | 11.8 [10.7, 13.0]              |
|   | <b>Anencephaly</b>                                     |                  |                  |                  |                  |                   | <b>185</b>                     |
|   | Q000-Q002  | 40               | 35               | 31               | 34               | 45                | 5.0 [4.3, 5.8]                 |
|   | <b>Encephalocele</b>                                   |                  |                  |                  |                  |                   | <b>50</b>                      |
|   | Q010-Q019  | 11               | 12               | 11               | 8                | 8                 | 1.4 [1.0, 1.8]                 |
|   | <b>Spina Bifida</b>                                    |                  |                  |                  |                  |                   | <b>214</b>                     |
|   | Q050-Q059  | 34               | 44               | 40               | 43               | 53                | 5.8 [5.1, 6.7]                 |
|   | <b>Isolated Hydrocephalus</b>                          |                  |                  |                  |                  |                   | <b>245</b>                     |
|   | Q030-Q039  | 37               | 67               | 46               | 58               | 37                | 6.7 [5.9, 7.6]                 |
| <b>Microcephaly</b>                                       |  |                  |                  |                  |                  | <b>ϕ</b>          |                                |
| Q02X  | 5  | 9                | <5               | 9                | <5               | 0.8 [0.5, 1.1]    |                                |
| <b>Eye anomalies</b>                                      |  |                  |                  |                  |                  | <b>58</b>         |                                |
| Q100-Q159   | 11   | 13               | 14               | 11               | 9                | 1.6 [1.2, 2.0]    |                                |
| <b>Ear anomalies (minor Q170-Q179 anomalies excluded)</b> |  |                  |                  |                  |                  | <b>ϕ</b>          |                                |
| Q160-Q169   | <5   | <5               | <5               | <5               | <5               | 0.3 [0.2, 0.6]    |                                |
| <b>CARDIOVASCULAR SYSTEM</b>                              | Q200-Q269  | 435              | 464              | 421              | 448              | 378               | <b>2146</b>                    |
|   |  | 62.0             | 64.0             | 56.5             | 60.5             | 49.9              | 58.5 [56.0, 61.0]              |
|   | <b>Serious cardiac#</b>                                |                  |                  |                  |                  |                   | <b>809</b>                     |
|   | Q200,Q203-4,Q212-3,Q220,<br>Q224-6,Q230,Q234,Q251,Q262 | 162              | 149              | 141              | 179              | 178               | 22.0 [20.6, 23.6]              |
|   | <b>Ventricular septal defect</b>                       |                  |                  |                  |                  |                   | <b>690</b>                     |
|   | Q210   | 141              | 141              | 158              | 129              | 121               | 18.8 [17.4, 20.3]              |
| <b>Atrial septal defect</b>                               |  |                  |                  |                  |                  | <b>459</b>        |                                |
| Q211  | 87   | 104              | 93               | 86               | 89               | 12.5 [11.4, 13.7] |                                |

|   |               | 2006             | 2007             | 2008             | 2009             | 2010             | TOTAL                          |
|---|---------------|------------------|------------------|------------------|------------------|------------------|--------------------------------|
|   |               | EMSYCAR births   |                  |                  |                  |                  |                                |
|   |               | 70153            | 72549            | 74469            | 74101            | 75698            | 366970                         |
|   | ICD-10        | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000<br>[95% C.I.] |
| <b>CARDIOVASCULAR SYSTEM (ctd.)</b>               |               |                  |                  |                  |                  |                  |                                |
| Atrio-ventricular septal defect                   | Q212          | 27<br>3.8        | 37<br>5.1        | 23<br>3.1        | 42<br>5.7        | 34<br>4.5        | 163<br>4.4 [3.8, 5.2]          |
| Fallot's Tetralogy                                | Q213          | 21<br>3.0        | 30<br>4.1        | 35<br>4.7        | 28<br>3.8        | 35<br>4.6        | 149<br>4.1 [3.4, 4.8]          |
| Transposition of the great vessels                | Q203          | 26<br>3.7        | 22<br>3.0        | 27<br>3.6        | 40<br>5.4        | 40<br>5.3        | 155<br>4.2 [3.6, 4.9]          |
| Pulmonary Valve Atresia                           | Q220          | 8<br>1.1         | 16<br>2.2        | 11<br>1.5        | 10<br>1.3        | 13<br>1.7        | 58<br>1.6 [1.2, 2.0]           |
| Pulmonary Valve Stenosis                          | Q221          | 20<br>2.9        | 19<br>2.6        | 16<br>2.1        | 21<br>2.8        | 27<br>3.6        | 103<br>2.8 [2.3, 3.4]          |
| Hypoplastic left heart syndrome                   | Q234          | 36<br>5.1        | 30<br>4.1        | 14<br>1.9        | 27<br>3.6        | 24<br>3.2        | 131<br>3.6 [3.0, 4.2]          |
| Coarctation of the aorta                          | Q251          | 27<br>3.8        | 23<br>3.2        | 33<br>4.4        | 27<br>3.6        | 28<br>3.7        | 138<br>3.8 [3.2, 4.4]          |
| Patent Ductus Arteriosus (>=37 weeks gest)        | Q250          | 56<br>8.0        | 64<br>8.8        | 47<br>6.3        | 38<br>5.1        | 42<br>5.5        | 247<br>6.7 [5.9, 7.6]          |
| <b>UROGENITAL SYSTEM</b>                          | Q500-Q649     | 370<br>52.7      | 372<br>51.3      | 340<br>45.7      | 299<br>40.4      | 213<br>28.1      | 1594<br>43.4 [41.3, 45.6]      |
| Renal agenesis and hypoplasia                     | Q600-Q609     | 34<br>4.8        | 41<br>5.7        | 32<br>4.3        | 30<br>4.0        | 19<br>2.5        | 156<br>4.3 [3.6, 5.0]          |
| Bilateral Renal Agenesis and/or Potters Syndrome# | Q601 and Q606 | 14<br>2.0        | 12<br>1.7        | 14<br>1.9        | 16<br>2.2        | 8<br>1.1         | 64<br>1.7 [1.3, 2.2]           |
| Bladder/urethral anomalies                        | Q640-Q649     | 27<br>3.8        | 25<br>3.4        | 37<br>5.0        | 28<br>3.8        | 25<br>3.3        | 142<br>3.9 [3.3, 4.6]          |
| Bladder exstrophy & Epispadias#                   | Q640 and Q641 | 6<br>0.9         | <5               | 8<br>1.1         | 5<br>0.7         | <5               | ϕ<br>0.7 [0.5, 1.0]            |
| Posterior Urethral Valves/Prune Belly Syndrome#   | Q6420 & Q794  | <5               | <5               | <5               | <5               | 8<br>1.1         | ϕ<br>0.7 [0.4, 1.0]            |
| Cystic kidneys                                    | Q610-Q619     | 38<br>5.4        | 48<br>6.6        | 63<br>8.5        | 42<br>5.7        | 44<br>5.8        | 235<br>6.4 [5.6, 7.3]          |



|   |               | <b>2006</b>           | <b>2007</b>                    | <b>2008</b>                    | <b>2009</b>                    | <b>2010</b>                    | <b>TOTAL</b>                   |   |
|---|---------------|-----------------------|--------------------------------|--------------------------------|--------------------------------|--------------------------------|--------------------------------|---|
|   |               | <b>EMSYCAR births</b> | 70153                          | 72549                          | 74469                          | 74101                          | 75698                          | <b>366970</b>                                       |
|   |               |                       | <i>n</i><br><i>Rate/10,000</i> | <i>n</i><br><i>Rate/10,000</i> | <i>n</i><br><i>Rate/10,000</i> | <i>n</i><br><i>Rate/10,000</i> | <i>n</i><br><i>Rate/10,000</i> | <i>n</i><br><i>Rate/10,000</i><br><i>[95% C.I.]</i> |
| <b>UROGENITAL SYSTEM (ctd.)</b>                     | <b>ICD-10</b> |                       |                                |                                |                                |                                |                                |   |
| Renal Dysplasia#                                    | Q614          | 28                    | 38                             | 47                             | 37                             | 37                             | 187                            |   |
|   |               | 4.0                   | 5.2                            | 6.3                            | 5.0                            | 4.9                            | 5.1 [4.4, 5.9]                 |   |
| Hypospadias and congenital chordee                  | Q540-Q549     | 126                   | 135                            | 99                             | 95                             | 65                             | 520                            |   |
|   |               | 18.0                  | 18.6                           | 13.3                           | 12.8                           | 8.6                            | 14.2 [13.0, 15.4]              |   |
| Hydronephrosis                                      | Q620          | 114                   | 106                            | 94                             | 88                             | 39                             | 441                            |   |
|   |               | 16.3                  | 14.6                           | 12.6                           | 11.9                           | 5.2                            | 12.0 [10.9, 13.2]              |   |
| <b>GASTRO-INTESTINAL SYSTEM</b>                     | Q350-Q459     | 200                   | 184                            | 214                            | 196                            | 200                            | 994                            |   |
|   |               | 28.5                  | 25.4                           | 28.7                           | 26.5                           | 26.4                           | 27.1 [25.4, 28.8]              |   |
| All oro-facial clefts                               | Q350-Q379     | 104                   | 98                             | 117                            | 87                             | 98                             | 504                            |   |
|   |               | 14.8                  | 13.5                           | 15.7                           | 11.7                           | 12.9                           | 13.7 [12.6, 15.0]              |   |
| Cleft palate only                                   | Q35           | 31                    | 30                             | 40                             | 33                             | 28                             | 162                            |   |
|   |               | 4.4                   | 4.1                            | 5.4                            | 4.5                            | 3.7                            | 4.4 [3.8, 5.1]                 |   |
| Cleft lip +/- Cleft Palate#                         | Q36 & Q37     | 73                    | 68                             | 78                             | 54                             | 70                             | 343                            |   |
|   |               | 10.4                  | 9.4                            | 10.5                           | 7.3                            | 9.2                            | 9.3 [8.4, 10.4]                |   |
| Atresia/stenosis small intestine                    | Q410-Q419     | 23                    | 18                             | 17                             | 24                             | 17                             | 99                             |   |
|   |               | 3.3                   | 2.5                            | 2.3                            | 3.2                            | 2.2                            | 2.7 [2.2, 3.3]                 |   |
| Duodenal atresia/stenosis#                          | Q410          | 14                    | 16                             | 12                             | 14                             | 6                              | 62                             |   |
|   |               | 2.0                   | 2.2                            | 1.6                            | 1.9                            | 0.8                            | 1.7 [1.3, 2.2]                 |   |
| Atresia/stenosis large intestine                    | Q420-Q429     | 26                    | 21                             | 33                             | 22                             | 30                             | 132                            |   |
|   |               | 3.7                   | 2.9                            | 4.4                            | 3.0                            | 4.0                            | 3.6 [3.0, 4.3]                 |   |
| Ano-rectal atresia/stenosis#                        | Q420-Q423     | 22                    | 17                             | 29                             | 18                             | 28                             | 114                            |   |
|   |               | 3.1                   | 2.3                            | 3.9                            | 2.4                            | 3.7                            | 3.1 [2.6, 3.7]                 |   |
| Other intestine                                     | Q430-Q439     | 27                    | 18                             | 33                             | 35                             | 42                             | 155                            |   |
|   |               | 3.8                   | 2.5                            | 4.4                            | 4.7                            | 5.5                            | 4.2 [3.6, 4.9]                 |   |
| Hirschsprung's disease#                             | Q431          | 8                     | <5                             | 9                              | 9                              | 13                             | ϕ                              |   |
|   |               | 1.1                   |                                | 1.2                            | 1.2                            | 1.7                            | 1.2 [0.8, 1.6]                 |   |
| Oesophageal atresia +/-Tracheo-Oesophageal fistula# | Q390-Q391     | 13                    | 14                             | 16                             | 20                             | 19                             | 82                             |   |
|   |               | 1.9                   | 1.9                            | 2.1                            | 2.7                            | 2.5                            | 2.2 [1.8, 2.8]                 |   |

| GASTROINTESTINAL SYSTEM (ctd.)             | ICD-10  | 2006             | 2007             | 2008             | 2009             | 2010             | TOTAL                     |                                |
|--|---|------------------|------------------|------------------|------------------|------------------|---------------------------|--------------------------------|
|  |   | EMSYCAR births   | 70153            | 72549            | 74469            | 74101            | 75698                     | 366970                         |
|  |   | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000          | n<br>Rate/10,000<br>[95% C.I.] |
| Congenital Diaphragmatic HerniaΩ           | Q790  | 24<br>3.4        | 23<br>3.2        | 27<br>3.6        | 27<br>3.6        | 22<br>2.9        | 123<br>3.4 [2.8, 4.0]     |                                |
| <b>MUSCULOSKELETAL SYSTEM</b>              | Q650-Q799, Q180-Q189,<br>Q380-Q389                                    | 455<br>64.9      | 452<br>62.3      | 447<br>60.0      | 431<br>58.2      | 362<br>47.8      | 2147<br>58.5 [56.1, 61.0] |                                |
| Limb                                       | Q650-<br>Q652,Q658,Q659,Q660,Q681,Q682,Q688,<br>Q69-Q749 (excl Q6281) | 324<br>46.2      | 308<br>42.5      | 295<br>39.6      | 291<br>39.3      | 240<br>31.7      | 1458<br>39.7 [37.7, 41.8] |                                |
| All Limb reductions                        | Q710-Q739   | 63               | 49               | 46               | 44               | 28               | 230                       |                                |
| Upper#                                     | Q71   | 9.0<br>36        | 6.8<br>31        | 6.2<br>32        | 5.9<br>29        | 3.7<br>17        | 6.3 [5.5, 7.1]<br>145     |                                |
| Lower#                                     | Q72   | 5.1<br>26        | 4.3<br>11        | 4.3<br>17        | 3.9<br>17        | 2.2<br>8         | 4.0 [3.3, 4.6]<br>79      |                                |
| Polydactyly                                | Q690-Q699   | 3.7<br>75        | 1.5<br>69        | 2.3<br>70        | 2.3<br>83        | 1.1<br>62        | 2.2 [1.7, 2.7]<br>359     |                                |
| Syndactyly                                 | Q700-Q709   | 10.7<br>48       | 9.5<br>48        | 9.4<br>38        | 11.2<br>46       | 8.2<br>34        | 9.8 [8.8, 10.8]<br>214    |                                |
| Skeletal Dysplasia#                        | Q7402,Q77,Q7800 & Q782-Q788   | 6.8<br>14        | 6.6<br>12        | 5.1<br>16        | 6.2<br>16        | 4.5<br>6         | 5.8 [5.1, 6.7]<br>64      |                                |
| Talipes equinovarus (other types excluded) | Q660  | 2.0<br>128       | 1.7<br>126       | 2.1<br>131       | 2.2<br>107       | 0.8<br>109       | 1.7 [1.3, 2.2]<br>601     |                                |
|  |   | 18.2             | 17.4             | 17.6             | 14.4             | 14.4             | 16.4 [15.1, 17.7]         |                                |
| <b>ABDOMINAL WALL DEFECTS#</b>             | Q792,Q793 & Q795  | 76<br>10.8       | 78<br>10.8       | 82<br>11.0       | 75<br>10.1       | 74<br>9.8        | 385<br>10.5 [9.5, 11.6]   |                                |
| Exomphalos                                 | Q792  | 31<br>4.4        | 36<br>5.0        | 35<br>4.7        | 33<br>4.5        | 26<br>3.4        | 161<br>4.4 [3.7, 5.1]     |                                |
| Gastroschisis                              | Q793  | 43<br>6.1        | 36<br>5.0        | 42<br>5.6        | 38<br>5.1        | 45<br>5.9        | 204<br>5.6 [4.8, 6.4]     |                                |
| <b>RESPIRATORY SYSTEM</b>                  | Q300-Q349   | 30<br>4.3        | 35<br>4.8        | 31<br>4.2        | 47<br>6.3        | 38<br>5.0        | 181<br>4.9 [4.2, 5.7]     |                                |

|   |                                     | 2006  | 2007             | 2008             | 2009             | 2010             | TOTAL                          |                       |
|---|-------------------------------------|---|------------------|------------------|------------------|------------------|--------------------------------|-----------------------|
|   |                                     | EMSYCAR births  |                  |                  |                  |                  |                                |                       |
|   |                                     | 70153   | 72549            | 74469            | 74101            | 75698            | 366970                         |                       |
|   |                                     | n<br>Rate/10,000  | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000 | n<br>Rate/10,000<br>[95% C.I.] |                       |
| RESPIRATORY SYSTEM (ctd.)                   | ICD-10                              |   |                  |                  |                  |                  |                                |                       |
| Congenital Cystic Adenomatoid Malformation# | Q3380                               | 9<br>1.3  | 13<br>1.8        | 8<br>1.1         | 15<br>2.0        | 14<br>1.8        | 59<br>1.6 [1.2, 2.1]           |                       |
| CHROMOSOMAL ANOMALIES                       | Q900-Q939 & Q960-Q999 (excl Q936)#  | 263<br>37.5   | 225<br>31.0      | 271<br>36.4      | 248<br>33.5      | 254<br>33.6      | 1261<br>34.4 [32.5, 36.3]      |                       |
| Trisomy 21*                                 | Q900-Q909                           | 132<br>18.8   | 116<br>16.0      | 147<br>19.7      | 134<br>18.1      | 123<br>16.2      | 652<br>17.8 [16.4, 19.2]       |                       |
| Trisomy 18*                                 | Q910-Q913                           | 40<br>5.7   | 27<br>3.7        | 44<br>5.9        | 40<br>5.4        | 47<br>6.2        | 198<br>5.4 [4.7, 6.2]          |                       |
| Trisomy 13*                                 | Q914-Q917                           | 20<br>2.9   | 19<br>2.6        | 15<br>2.0        | 18<br>2.4        | 24<br>3.2        | 96<br>2.6 [2.1, 3.2]           |                       |
| Turner's Syndrome*                          | Q960-Q969                           | 17<br>2.4   | 13<br>1.8        | 27<br>3.6        | 21<br>2.8        | 28<br>3.7        | 106<br>2.9 [2.4, 3.5]          |                       |
| All other chromosomes (excluding * above)   | Q920-Q939<br>&Q970-Q999             | 54<br>7.7   | 50<br>6.9        | 38<br>5.1        | 35<br>4.7        | 32<br>4.2        | 209<br>5.7 [4.9, 6.5]          |                       |
| SYNDROMES AFFECTING MULTIPLE SYSTEMS        | Q870-Q879                           | 35<br>5.0   | 38<br>5.2        | 23<br>3.1        | 27<br>3.6        | 26<br>3.4        | 149<br>4.1 [3.4, 4.8]          |                       |
| SKIN & INTEGUMENT                           | Q800 - Q849                         | 10<br>1.4   | 11<br>1.5        | 11<br>1.5        | 5<br>0.7         | 5<br>0.7         | 42<br>1.1 [0.8, 1.5]           |                       |
| OTHER ANOMALIES                             | Isolated Cystic Hygroma             | D1810   | 9<br>1.3         | 20<br>2.8        | 14<br>1.9        | 15<br>2.0        | 19<br>2.5                      | 77<br>2.1 [1.7, 2.6]  |
|   | Other Cystic Hygroma                | D1810 (+ other anomalies)   | 35<br>5.0        | 36<br>5.0        | 44<br>5.9        | 47<br>6.3        | 55<br>7.3                      | 217<br>5.2, 6.8       |
|   | Genetic Syndromes & Microdeletions# | D821,Q4471,Q6190,Q7484,Q751,Q754,Q7581,<br>Q936,Q87 (but not Q8793,Q8704,Q8706,Q8708,<br>Q8716, Q8724 or Q8726) | 27<br>3.8        | 31<br>4.3        | 20<br>2.7        | 21<br>2.8        | 19<br>2.5                      | 118<br>2.7, 3.9       |
|   | Endocrine & Metabolic               | E000-E359 & E700-E90**  | 69<br>9.8        | 21<br>2.9        | 20<br>2.7        | 19<br>2.6        | 15<br>2.0                      | 144<br>3.9 [3.3, 4.6] |
|   | Other anomalies                     | Q851-Q869; Q890-Q899; D550; D573#   | 32<br>4.6        | 26<br>3.6        | 10<br>1.3        | 16<br>2.2        | 19<br>2.5                      | 103<br>2.8 [2.3, 3.4] |

# New subgroup included or change of definition (EUROCAT)

\*\* Data no longer routinely received from Neonatal Screening

Ω Subgroup moved      φ Small numbers suppressed

**Table 6. Anomalies by Birth Status 2006-2010: % by Anomaly Type**

|   | TOP<br>% | Fetal Loss<br>% | Stillbirth<br>% | Livebirth<br>% |
|---|----------|-----------------|-----------------|----------------|
| <b>All EMSYCAR cases</b>                          | 18.9     | 2.1             | 2.0             | 77.0           |
| <b>All Central Nervous System</b>                 | 57.6     | 2.1             | 4.1             | 36.2           |
| All Neural Tube Defects                           | 73.4     | 2.3             | 2.5             | 21.7           |
| Anencephaly                                       | 83.8     | 3.2             | 2.7             | 10.3           |
| Encephalocele                                     | 70.0     | 6.0             | 4.0             | 20.0           |
| Spina Bifida                                      | 65.4     | 0.9             | 1.9             | 31.8           |
| <b>All Cardiovascular</b>                         | 10.8     | 1.2             | 2.5             | 85.5           |
| Serious cardiac anomalies                         | 16.1     | 1.1             | 2.7             | 80.1           |
| Ventricular Septal Defect                         | 7.0      | 0.7             | 2.2             | 90.1           |
| Atrial Septal Defect (excl. Patent Foramen Ovale) | 3.1      | 0.7             | 0.7             | 95.6           |
| Transposition of the Great Arteries               | 10.3     | 0.0             | 1.9             | 87.7           |
| Hypoplastic Left Heart                            | 38.9     | 1.5             | 2.3             | 57.3           |
| Tetralogy of Fallot                               | 9.4      | 2.0             | 1.3             | 87.2           |
| Atrio-ventricular Septal Defect                   | 14.1     | 0.6             | 4.9             | 80.4           |
| Co-arctation of the Aorta                         | 2.2      | 1.4             | 1.4             | 94.9           |
| <b>All Respiratory</b>                            | 13.8     | 1.7             | 2.2             | 82.3           |
| Congenital Cystic Adenomatoid Malformation        | 5.1      | 1.7             | 1.7             | 91.5           |
| <b>All Gastro-intestinal</b>                      | 9.7      | 1.0             | 2.1             | 87.2           |
| Tracheo-oesophageal fistula +/- atresia           | 2.4      | 0.0             | 3.7             | 93.9           |
| All oro-facial clefts                             | 11.9     | 0.8             | 1.4             | 85.9           |
| Cleft Palate                                      | 1.9      | 0.0             | 1.2             | 96.9           |
| Cleft Lip +/- Palate                              | 16.6     | 1.2             | 1.5             | 80.8           |
| Diaphragmatic Hernia                              | 21.1     | 1.6             | 4.9             | 72.4           |
| <b>All Urogenital</b>                             | 9.6      | 1.1             | 1.5             | 87.8           |
| Renal Agenesis                                    | 41.7     | 2.6             | 5.1             | 50.6           |
| Cystic Kidneys                                    | 18.7     | 0.4             | 2.6             | 78.3           |
| Hydronephrosis                                    | 2.7      | 0.2             | 0.2             | 96.8           |
| Bladder/urethral                                  | 33.8     | 4.9             | 0.7             | 60.6           |
| <b>All Musculoskeletal</b>                        | 15.7     | 2.4             | 2.4             | 79.5           |
| Limb Reductions                                   | 23.5     | 3.0             | 3.5             | 70.0           |
| Skeletal Dysplasia                                | 51.6     | 0.0             | 4.7             | 43.8           |
| Gastroschisis                                     | 7.4      | 3.9             | 2.0             | 86.8           |
| Exomphalos  | 52.2     | 13.7            | 4.3             | 29.8           |
| <b>All Syndromes</b>                              | 20.1     | 2.0             | 5.4             | 72.5           |
| <b>All Chromosome</b>                             | 50.5     | 5.3             | 3.4             | 40.8           |
| Down Syndrome (Trisomy 21)                        | 47.4     | 3.1             | 1.5             | 48.0           |
| Edwards Syndrome (Trisomy 18)                     | 68.2     | 5.1             | 9.1             | 17.7           |
| Patau Syndrome (Trisomy 13)                       | 72.9     | 5.2             | 4.2             | 17.7           |
| Turner Syndrome (45X)                             | 55.7     | 20.8            | 0.9             | 22.6           |

**Table 7. Anomalies by Maternal Age 2006 – 2010: rates per 10,000 births**

|   | <20yrs       | 20-24        | 25-29        | 30-34        | 35-39        | > 40yrs      | Age NK*     | Age NK Proportion<br>EMSYCAR cases |
|---|--------------|--------------|--------------|--------------|--------------|--------------|-------------|------------------------------------|
|   | per 10,000   | per 10,000   | per 10,000   | per 10,000   | per 10,000   | per 10,000   | per 10,000  | %                                  |
| <b>All EMSYCAR cases</b>                          | <b>224.8</b> | <b>192.8</b> | <b>187.0</b> | <b>191.1</b> | <b>231.1</b> | <b>403.2</b> | <b>17.4</b> | <b>7.8</b>                         |
| <b>All Central Nervous System</b>                 | <b>30.4</b>  | <b>23.0</b>  | <b>22.0</b>  | <b>20.0</b>  | <b>25.4</b>  | <b>25.3</b>  | <b>0.5</b>  | <b>2.2</b>                         |
| All Neural Tube Defects                           | 15.5         | 11.1         | 10.7         | 10.7         | 13.6         | 13.1         | 0.2         | 1.4                                |
| Anencephaly                                       | 7.1          | 5.4          | 3.9          | 4.3          | 6.1          | 7.0          | 0.1         | 1.1                                |
| Encephalocele                                     | 1.1          | 0.7          | 2.2          | 1.1          | 1.0          | 3.5          | 0.0         | 0.0                                |
| Spina Bifida                                      | 8.5          | 5.4          | 5.2          | 5.3          | 6.7          | 4.4          | 0.1         | 2.3                                |
| <b>All Cardiovascular</b>                         | <b>44.5</b>  | <b>39.8</b>  | <b>42.5</b>  | <b>45.7</b>  | <b>54.9</b>  | <b>78.5</b>  | <b>12.7</b> | <b>21.7</b>                        |
| Serious cardiac                                   | 19.1         | 14.4         | 16.9         | 18.7         | 20.6         | 26.2         | 4.3         | 19.3                               |
| Ventricular Septal Defect                         | 12.7         | 13.7         | 14.6         | 15.2         | 19.1         | 33.2         | 3.2         | 16.8                               |
| Atrial Septal Defect (excl. Patent Foramen Ovale) | 8.1          | 9.7          | 9.6          | 9.1          | 9.0          | 21.8         | 2.8         | 22.7                               |
| Transposition of the Great Arteries               | 3.9          | 2.9          | 3.2          | 4.1          | 2.7          | 7.9          | 0.7         | 17.4                               |
| Hypoplastic Left Heart                            | 4.9          | 3.0          | 3.2          | 3.1          | 4.2          | 1.7          | 0.2         | 5.3                                |
| Tetralogy of Fallot                               | 4.9          | 2.6          | 3.1          | 2.9          | 3.4          | 1.7          | 1.0         | 23.5                               |
| Atrio-ventricular Septal Defect                   | 2.8          | 2.4          | 3.3          | 2.7          | 6.9          | 10.5         | 0.8         | 17.8                               |
| Co-arctation of the Aorta                         | 2.1          | 2.9          | 2.0          | 3.4          | 2.7          | 5.2          | 1.0         | 27.5                               |
| <b>All Respiratory</b>                            | <b>3.5</b>   | <b>4.5</b>   | <b>4.3</b>   | <b>3.9</b>   | <b>3.6</b>   | <b>7.0</b>   | <b>0.8</b>  | <b>16.4</b>                        |
| Congenital Cystic Adenomatoid Malformation        | 2.1          | 1.7          | 1.9          | 1.4          | 1.0          | 0.9          | 0.0         | 1.9                                |
| <b>All Gastro-intestinal</b>                      | <b>26.8</b>  | <b>25.3</b>  | <b>24.8</b>  | <b>25.4</b>  | <b>22.8</b>  | <b>38.4</b>  | <b>1.7</b>  | <b>6.3</b>                         |
| Tracheo-oesophageal Fistula +/- atresia           | 1.8          | 1.1          | 1.9          | 2.6          | 2.3          | 3.5          | 0.2         | 13.2                               |
| All oro-facial clefts                             | 16.2         | 14.1         | 13.3         | 13.4         | 12.0         | 15.7         | 0.1         | 1.2                                |
| Cleft Palate                                      | 2.8          | 4.5          | 4.6          | 4.8          | 3.8          | 3.5          | 0.1         | 2.5                                |
| Cleft Lip +/- Palate                              | 13.4         | 9.6          | 8.7          | 8.6          | 8.4          | 12.2         | 0.0         | 0.4                                |
| Congenital Diaphragmatic Hernia                   | 3.2          | 2.5          | 3.5          | 2.4          | 5.0          | 3.5          | 0.2         | 6.4                                |

**Table 7. ctd. Anomalies by Maternal Age**

|                            | <20yrs      | 20-24       | 25-29       | 30-34       | 35-39       | > 40yrs      | Age NK*    | Age NK Proportion<br>EMSYCAR cases |
|----------------------------|-------------|-------------|-------------|-------------|-------------|--------------|------------|------------------------------------|
|                            | per 10,000  | per 10,000  | per 10,000  | per 10,000  | per 10,000  | per 10,000   | per 10,000 | %                                  |
| <b>All urogenital</b>      | <b>42.7</b> | <b>46.8</b> | <b>41.6</b> | <b>41.5</b> | <b>43.8</b> | <b>40.1</b>  | <b>0.4</b> | <b>0.9</b>                         |
| Renal Agenesis             | 6.0         | 4.7         | 3.8         | 3.9         | 4.2         | 3.5          | 0.0        | 0.0                                |
| Cystic Kidneys             | 5.3         | 7.2         | 6.0         | 6.7         | 6.1         | 4.4          | 0.1        | 0.9                                |
| Hydronephrosis             | 15.9        | 13.0        | 10.8        | 10.6        | 13.2        | 13.1         | 0.0        | 0.0                                |
| Bladder/urethral           | 2.8         | 4.1         | 3.4         | 4.2         | 4.0         | 3.5          | 0.1        | 1.4                                |
| <b>All Musculoskeletal</b> | <b>82.9</b> | <b>55.7</b> | <b>56.6</b> | <b>50.7</b> | <b>58.9</b> | <b>76.8</b>  | <b>0.6</b> | <b>1.0</b>                         |
| Limb Reductions            | 7.4         | 5.0         | 6.9         | 5.8         | 5.9         | 11.3         | 0.0        | 0.0                                |
| Skeletal Dysplasia         | 1.4         | 1.7         | 1.6         | 1.7         | 2.3         | 1.7          | 0.0        | 0.0                                |
| Gastroschisis              | 28.9        | 8.6         | 3.2         | 1.3         | 0.8         | 3.5          | 0.0        | 0.0                                |
| Exomphalos                 | 4.6         | 3.0         | 3.6         | 3.5         | 6.5         | 18.3         | 0.0        | 0.0                                |
| <b>All Syndromes</b>       | <b>2.1</b>  | <b>4.2</b>  | <b>3.7</b>  | <b>3.7</b>  | <b>4.2</b>  | <b>7.0</b>   | <b>0.2</b> | <b>4.7</b>                         |
| <b>All Chromosome</b>      | <b>19.1</b> | <b>15.0</b> | <b>18.5</b> | <b>28.1</b> | <b>68.1</b> | <b>221.7</b> | <b>1.2</b> | <b>3.5</b>                         |
| Trisomy 21                 | 6.4         | 6.1         | 7.4         | 13.0        | 41.7        | 134.4        | 0.5        | 2.6                                |
| Trisomy 18                 | 2.8         | 2.1         | 2.2         | 4.3         | 9.8         | 50.6         | 0.1        | 1.0                                |
| Trisomy 13                 | 0.7         | 0.5         | 1.8         | 2.8         | 6.1         | 12.2         | 0.0        | 0.0                                |
| 45X                        | 6.0         | 2.2         | 2.2         | 2.6         | 3.1         | 3.5          | 0.1        | 3.8                                |

\* proportion of all births

**Table 8. Anomalies by PCT 2006-2010: rates per 10,000 births****Barnsley**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 10.9  | 39.8 | 39.4 | 14.1 | 30.9 | <b>27.0</b> | [19.1, 37.1] |
| Eye, ear               | Q100-Q179                       | 0.0   | 0.0  | 0.0  | 3.5  | 3.4  | <b>1.4</b>  | -            |
| Cardiovascular         | Q200-Q269                       | 32.6  | 32.5 | 25.1 | 45.8 | 68.7 | <b>41.2</b> | [31.3, 53.3] |
| Serious Cardiac        | Q20 various (see Table 10)      | 21.7  | 18.1 | 14.3 | 24.7 | 24.0 | <b>20.6</b> | [13.8, 29.6] |
| Urogenital             | Q500-Q649                       | 36.2  | 21.7 | 39.4 | 21.1 | 24.0 | <b>28.4</b> | [20.3, 38.7] |
| Gastrointestinal       | Q350-Q459                       | 25.4  | 25.3 | 35.8 | 24.7 | 58.4 | <b>34.1</b> | [25.2, 45.2] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 47.1  | 68.7 | 53.8 | 56.4 | 68.7 | <b>59.0</b> | [47.0, 73.1] |
| Respiratory            | A300-Q349                       | 0.0   | 0.0  | 3.6  | 7.0  | 6.9  | <b>3.6</b>  | [1.2, 8.3]   |
| Chromosomal            | Q900-Q999                       | 25.4  | 21.7 | 43.0 | 17.6 | 27.5 | <b>27.0</b> | [19.1, 37.1] |
| Syndromes              | Q870-Q879                       | 3.6   | 3.6  | 0.0  | 0.0  | 0.0  | <b>1.4</b>  | -            |

**Bassetlaw**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 51.4  | 23.9 | 24.9 | 25.2 | 46.8 | <b>34.4</b> | [21.3, 52.6] |
| Eye, ear               | Q100-Q179                       | 0.0   | 0.0  | 0.0  | 0.0  | 0.0  | <b>0.0</b>  | -            |
| Cardiovascular         | Q200-Q269                       | 34.3  | 16.0 | 58.0 | 42.0 | 23.4 | <b>34.4</b> | [21.3, 52.6] |
| Serious Cardiac        | Q20 various (see Table 10)      | 17.1  | 8.0  | 33.2 | 8.4  | 7.8  | <b>14.8</b> | [6.7, 28.0]  |
| Urogenital             | Q500-Q649                       | 25.7  | 8.0  | 16.6 | 16.8 | 0.0  | <b>13.1</b> | [5.7, 25.8]  |
| Gastrointestinal       | Q350-Q459                       | 17.1  | 55.9 | 49.8 | 33.6 | 39.0 | <b>39.4</b> | [25.2, 58.5] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 51.4  | 39.9 | 33.2 | 50.4 | 62.4 | <b>47.5</b> | [31.9, 68.2] |
| Respiratory            | A300-Q349                       | 0.0   | 8.0  | 8.3  | 0.0  | 7.8  | <b>4.9</b>  | -            |
| Chromosomal            | Q900-Q999                       | 25.7  | 31.9 | 66.3 | 42.0 | 39.0 | <b>41.0</b> | [26.5, 60.5] |
| Syndromes              | Q870-Q879                       | 0.0   | 8.0  | 8.3  | 8.4  | 7.8  | <b>6.6</b>  | [1.8, 16.8]  |

**Derby City**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 18.2  | 20.9 | 2.7  | 13.9 | 22.6 | <b>15.5</b> | [10.2, 22.5] |
| Eye, ear               | Q100-Q179                       | 0.0   | 0.0  | 0.0  | 0.0  | 0.0  | <b>0.0</b>  | -            |
| Cardiovascular         | Q200-Q269                       | 76.0  | 56.7 | 38.5 | 55.6 | 36.7 | <b>52.2</b> | [42.1, 64.1] |
| Serious Cardiac        | Q20 various (see Table 10)      | 36.5  | 23.9 | 11.0 | 27.8 | 19.8 | <b>23.5</b> | [16.9, 31.9] |
| Urogenital             | Q500-Q649                       | 85.2  | 44.7 | 52.2 | 36.1 | 25.4 | <b>48.2</b> | [38.5, 59.7] |
| Gastrointestinal       | Q350-Q459                       | 21.3  | 23.9 | 38.5 | 27.8 | 28.2 | <b>28.1</b> | [20.8, 37.2] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 100.4 | 56.7 | 74.2 | 52.8 | 53.7 | <b>67.2</b> | [55.6, 80.4] |
| Respiratory            | A300-Q349                       | 6.1   | 6.0  | 0.0  | 2.8  | 2.8  | <b>3.4</b>  | [1.3, 7.5]   |
| Chromosomal            | Q900-Q999                       | 24.3  | 17.9 | 43.9 | 19.5 | 31.1 | <b>27.6</b> | [20.3, 36.5] |
| Syndromes              | Q870-Q879                       | 6.1   | 0.0  | 2.7  | 2.8  | 0.0  | <b>2.3</b>  | [0.6, 5.9]   |



**Table 8 ctd. Anomalies by PCT 2006-2010: rates per 10,000 births**

**Derbyshire County**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 19.7  | 19.3 | 18.9 | 20.7 | 22.3 | <b>20.2</b> | [16.0, 25.2] |
| Eye, ear               | Q100-Q179                       | 0.0   | 1.3  | 1.3  | 3.9  | 1.2  | <b>1.5</b>  | [0.6, 3.3]   |
| Cardiovascular         | Q200-Q269                       | 44.6  | 41.3 | 32.7 | 46.5 | 37.2 | <b>40.4</b> | [34.3, 47.2] |
| Serious Cardiac        | Q20 various (see Table 10)      | 27.6  | 20.6 | 10.1 | 21.9 | 18.6 | <b>19.7</b> | [15.5, 24.6] |
| Urogenital             | Q500-Q649                       | 42.0  | 38.7 | 22.6 | 28.4 | 17.4 | <b>29.6</b> | [24.5, 35.5] |
| Gastrointestinal       | Q350-Q459                       | 24.9  | 16.8 | 27.7 | 24.5 | 27.3 | <b>24.3</b> | [19.6, 29.7] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 55.1  | 41.3 | 42.7 | 42.6 | 45.9 | <b>45.5</b> | [39.1, 52.7] |
| Respiratory            | A300-Q349                       | 0.0   | 0.0  | 0.0  | 6.5  | 3.7  | <b>2.0</b>  | [0.9, 4.0]   |
| Chromosomal            | Q900-Q999                       | 27.6  | 29.7 | 28.9 | 31.0 | 16.1 | <b>26.6</b> | [21.7, 32.2] |
| Syndromes              | Q870-Q879                       | 5.2   | 3.9  | 1.3  | 1.3  | 3.7  | <b>3.1</b>  | [1.6, 5.4]   |

**Doncaster**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |               |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|---------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.      |
| Central Nervous System | Q000-Q079                       | 30.3  | 29.7 | 28.8 | 31.1 | 15.7 | <b>27.1</b> | [27.0, 47.6]  |
| Eye, ear               | Q100-Q179                       | 0.0   | 0.0  | 2.6  | 0.0  | 0.0  | <b>0.5</b>  | -             |
| Cardiovascular         | Q200-Q269                       | 30.3  | 27.0 | 47.1 | 25.9 | 31.5 | <b>32.4</b> | [33.2, 55.7]  |
| Serious Cardiac        | Q20 various (see Table 10)      | 16.5  | 10.8 | 23.5 | 7.8  | 15.7 | <b>14.9</b> | [13.2, 28.8]  |
| Urogenital             | Q500-Q649                       | 46.9  | 37.9 | 47.1 | 33.7 | 36.7 | <b>40.4</b> | [42.6, 67.6]  |
| Gastrointestinal       | Q350-Q459                       | 66.2  | 35.2 | 26.1 | 28.5 | 21.0 | <b>35.1</b> | [36.3, 59.6]  |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 68.9  | 59.5 | 73.2 | 62.2 | 65.5 | <b>65.9</b> | [73.4, 105.0] |
| Respiratory            | A300-Q349                       | 5.5   | 2.7  | 7.8  | 2.6  | 0.0  | <b>3.7</b>  | [2.0, 10.2]   |
| Chromosomal            | Q900-Q999                       | 24.8  | 16.2 | 54.9 | 28.5 | 47.2 | <b>34.5</b> | [35.7, 58.9]  |
| Syndromes              | Q870-Q879                       | 0.0   | 2.7  | 2.6  | 5.2  | 2.6  | <b>2.7</b>  | -             |

**Leicester City**

| Anomaly Group          | ICD-10                          | Rates |       |       |      |       | TOTAL        |               |
|------------------------|---------------------------------|-------|-------|-------|------|-------|--------------|---------------|
|                        |                                 | 2006  | 2007  | 2008  | 2009 | 2010  | Rate         | 95% C.I.      |
| Central Nervous System | Q000-Q079                       | 29.2  | 37.5  | 40.3  | 28.6 | 33.7  | <b>33.9</b>  | [27.2, 41.8]  |
| Eye, ear               | Q100-Q179                       | 0.0   | 7.9   | 5.8   | 3.8  | 7.5   | <b>5.1</b>   | [2.7, 8.7]    |
| Cardiovascular         | Q200-Q269                       | 62.6  | 130.3 | 113.1 | 91.5 | 118.0 | <b>103.7</b> | [91.6, 116.8] |
| Serious Cardiac        | Q20 various (see Table 10)      | 20.9  | 31.6  | 26.8  | 26.7 | 33.7  | <b>28.1</b>  | [22.0, 35.3]  |
| Urogenital             | Q500-Q649                       | 112.7 | 80.9  | 82.5  | 99.1 | 74.9  | <b>89.6</b>  | [78.5, 101.9] |
| Gastrointestinal       | Q350-Q459                       | 20.9  | 31.6  | 53.7  | 36.2 | 33.7  | <b>35.5</b>  | [28.6, 43.5]  |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 116.9 | 71.1  | 70.9  | 95.3 | 54.3  | <b>81.1</b>  | [70.5, 92.8]  |
| Respiratory            | A300-Q349                       | 6.3   | 3.9   | 7.7   | 7.6  | 5.6   | <b>6.2</b>   | [3.6, 10.1]   |
| Chromosomal            | Q900-Q999                       | 41.8  | 45.4  | 47.9  | 38.1 | 52.4  | <b>45.2</b>  | [37.4, 54.2]  |
| Syndromes              | Q870-Q879                       | 8.4   | 2.0   | 7.7   | 7.6  | 7.5   | <b>6.6</b>   | [3.9, 10.6]   |

**Table 8 ctd. Anomalies by PCT 2006-2010: rates per 10,000 births**

**Leicestershire County & Rutland**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 25.4  | 24.9 | 27.7 | 41.6 | 17.4 | <b>27.3</b> | [22.2, 33.3] |
| Eye, ear               | Q100-Q179                       | 7.0   | 5.5  | 1.4  | 2.8  | 4.0  | <b>4.1</b>  | [2.3, 6.8]   |
| Cardiovascular         | Q200-Q269                       | 78.9  | 70.6 | 69.2 | 95.8 | 61.7 | <b>75.1</b> | [66.5, 84.6] |
| Serious Cardiac        | Q20 various (see Table 10)      | 26.8  | 20.8 | 20.8 | 26.4 | 14.8 | <b>21.8</b> | [17.3, 27.2] |
| Urogenital             | Q500-Q649                       | 74.7  | 85.8 | 54.0 | 41.6 | 52.3 | <b>61.6</b> | [53.8, 70.2] |
| Gastrointestinal       | Q350-Q459                       | 29.6  | 38.7 | 31.9 | 26.4 | 22.8 | <b>29.8</b> | [24.5, 36.0] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 62.0  | 80.3 | 66.5 | 59.7 | 41.6 | <b>61.9</b> | [54.1, 70.5] |
| Respiratory            | A300-Q349                       | 5.6   | 8.3  | 4.2  | 11.1 | 12.1 | <b>8.3</b>  | [5.6, 11.8]  |
| Chromosomal            | Q900-Q999                       | 73.3  | 54.0 | 62.3 | 36.1 | 52.3 | <b>55.5</b> | [48.1, 63.7] |
| Syndromes              | Q870-Q879                       | 4.2   | 5.5  | 0.0  | 2.8  | 4.0  | <b>3.3</b>  | [1.7, 5.8]   |

**Lincolnshire**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 13.1  | 25.0 | 10.3 | 20.9 | 16.6 | <b>17.2</b> | [13.2, 21.9] |
| Eye, ear               | Q100-Q179                       | 0.0   | 4.2  | 0.0  | 1.3  | 0.0  | <b>1.1</b>  | [0.3, 2.7]   |
| Cardiovascular         | Q200-Q269                       | 48.0  | 37.5 | 47.8 | 56.1 | 34.6 | <b>44.8</b> | [38.3, 52.1] |
| Serious Cardiac        | Q20 various (see Table 10)      | 20.3  | 16.7 | 12.9 | 23.5 | 23.1 | <b>19.3</b> | [15.1, 24.3] |
| Urogenital             | Q500-Q649                       | 20.3  | 26.4 | 32.3 | 37.8 | 11.5 | <b>25.7</b> | [20.9, 31.4] |
| Gastrointestinal       | Q350-Q459                       | 29.1  | 20.8 | 32.3 | 24.8 | 14.1 | <b>24.1</b> | [19.4, 29.6] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 52.3  | 43.0 | 43.9 | 43.0 | 38.4 | <b>44.0</b> | [37.5, 51.2] |
| Respiratory            | A300-Q349                       | 0.0   | 5.6  | 2.6  | 5.2  | 2.6  | <b>3.2</b>  | [1.7, 5.6]   |
| Chromosomal            | Q900-Q999                       | 39.2  | 33.3 | 28.4 | 37.8 | 32.0 | <b>34.0</b> | [28.4, 40.5] |
| Syndromes              | Q870-Q879                       | 1.5   | 5.6  | 3.9  | 3.9  | 0.0  | <b>2.9</b>  | [1.5, 5.3]   |

**North East Lincolnshire**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 15.3  | 39.9 | 5.1  | 30.0 | 34.9 | <b>25.1</b> | [16.3, 37.0] |
| Eye, ear               | Q100-Q179                       | 0.0   | 5.0  | 5.1  | 5.0  | 5.0  | <b>4.0</b>  | [1.1, 10.3]  |
| Cardiovascular         | Q200-Q269                       | 20.4  | 49.9 | 65.7 | 79.9 | 29.9 | <b>49.2</b> | [36.4, 65.0] |
| Serious Cardiac        | Q20 various (see Table 10)      | 15.3  | 24.9 | 10.1 | 25.0 | 14.9 | <b>18.1</b> | [10.7, 28.6] |
| Urogenital             | Q500-Q649                       | 56.0  | 44.9 | 45.5 | 54.9 | 14.9 | <b>43.2</b> | [31.3, 58.1] |
| Gastrointestinal       | Q350-Q459                       | 56.0  | 44.9 | 55.6 | 69.9 | 39.9 | <b>53.2</b> | [39.9, 69.6] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 76.4  | 79.8 | 30.3 | 99.9 | 34.9 | <b>64.3</b> | [49.5, 82.0] |
| Respiratory            | A300-Q349                       | 0.0   | 10.0 | 10.1 | 0.0  | 10.0 | <b>6.0</b>  | [2.2, 13.1]  |
| Chromosomal            | Q900-Q999                       | 40.8  | 10.0 | 20.2 | 25.0 | 24.9 | <b>24.1</b> | [15.5, 35.9] |
| Syndromes              | Q870-Q879                       | 5.1   | 0.0  | 0.0  | 15.0 | 0.0  | <b>4.0</b>  | [1.1, 10.3]  |

**Table 8 ctd. Anomalies by PCT 2006-2010: rates per 10,000 births****North Lincolnshire**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 11.0  | 16.2 | 16.1 | 15.7 | 15.9 | <b>15.0</b> | [8.2, 25.2]  |
| Eye, ear               | Q100-Q179                       | 5.5   | 0.0  | 0.0  | 0.0  | 10.6 | <b>3.2</b>  | -            |
| Cardiovascular         | Q200-Q269                       | 77.0  | 43.3 | 21.5 | 26.2 | 42.3 | <b>41.8</b> | [29.7, 57.1] |
| Serious Cardiac        | Q20 various (see Table 10)      | 27.5  | 27.1 | 5.4  | 20.9 | 26.4 | <b>21.4</b> | [13.1, 33.1] |
| Urogenital             | Q500-Q649                       | 49.5  | 37.9 | 53.7 | 47.1 | 31.7 | <b>43.9</b> | [31.6, 59.6] |
| Gastrointestinal       | Q350-Q459                       | 38.5  | 21.7 | 21.5 | 36.6 | 26.4 | <b>28.9</b> | [19.1, 42.1] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 49.5  | 86.6 | 80.6 | 83.8 | 58.1 | <b>71.8</b> | [55.7, 91.1] |
| Respiratory            | A300-Q349                       | 5.5   | 0.0  | 0.0  | 0.0  | 0.0  | <b>1.1</b>  | -            |
| Chromosomal            | Q900-Q999                       | 11.0  | 21.7 | 32.2 | 5.2  | 21.1 | <b>18.2</b> | [10.6, 29.2] |
| Syndromes              | Q870-Q879                       | 11.0  | 16.2 | 5.4  | 5.2  | 10.6 | <b>9.6</b>  | [4.4, 18.3]  |

**Northamptonshire**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 19.7  | 16.6 | 21.6 | 20.9 | 27.9 | <b>21.4</b> | [17.3, 26.1] |
| Eye, ear               | Q100-Q179                       | 1.2   | 0.0  | 2.2  | 1.1  | 0.0  | <b>0.9</b>  | [0.2, 2.3]   |
| Cardiovascular         | Q200-Q269                       | 47.5  | 33.2 | 38.8 | 44.0 | 52.6 | <b>43.2</b> | [37.4, 49.7] |
| Serious Cardiac        | Q20 various (see Table 10)      | 26.6  | 11.1 | 18.3 | 19.8 | 32.2 | <b>21.6</b> | [17.5, 26.3] |
| Urogenital             | Q500-Q649                       | 40.5  | 36.5 | 36.7 | 23.1 | 17.2 | <b>30.6</b> | [25.8, 36.2] |
| Gastrointestinal       | Q350-Q459                       | 18.5  | 18.8 | 12.9 | 15.4 | 10.7 | <b>15.2</b> | [11.8, 19.2] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 45.1  | 47.6 | 37.7 | 37.4 | 44.0 | <b>42.3</b> | [36.6, 48.7] |
| Respiratory            | A300-Q349                       | 4.6   | 3.3  | 3.2  | 5.5  | 6.4  | <b>4.6</b>  | [2.9, 7.1]   |
| Chromosomal            | Q900-Q999                       | 26.6  | 16.6 | 16.2 | 35.2 | 30.1 | <b>24.9</b> | [20.5, 29.9] |
| Syndromes              | Q870-Q879                       | 6.9   | 6.6  | 2.2  | 3.3  | 2.1  | <b>4.2</b>  | [2.5, 6.5]   |

**Nottingham City**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 33.1  | 21.8 | 19.1 | 18.4 | 15.5 | <b>21.3</b> | [15.6, 28.5] |
| Eye, ear               | Q100-Q179                       | 5.1   | 0.0  | 2.4  | 0.0  | 0.0  | <b>1.4</b>  | -            |
| Cardiovascular         | Q200-Q269                       | 58.5  | 46.0 | 42.9 | 46.0 | 42.2 | <b>46.9</b> | [38.1, 57.1] |
| Serious Cardiac        | Q20 various (see Table 10)      | 20.4  | 21.8 | 33.3 | 23.0 | 26.6 | <b>25.1</b> | [18.8, 32.8] |
| Urogenital             | Q500-Q649                       | 76.4  | 55.7 | 61.9 | 46.0 | 28.9 | <b>53.1</b> | [43.7, 63.8] |
| Gastrointestinal       | Q350-Q459                       | 15.3  | 9.7  | 21.4 | 29.9 | 17.8 | <b>19.0</b> | [13.5, 25.8] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 89.1  | 65.4 | 78.6 | 71.3 | 88.8 | <b>78.7</b> | [67.2, 91.5] |
| Respiratory            | A300-Q349                       | 5.1   | 4.8  | 2.4  | 16.1 | 4.4  | <b>6.6</b>  | [3.6, 11.1]  |
| Chromosomal            | Q900-Q999                       | 45.8  | 24.2 | 38.1 | 29.9 | 39.9 | <b>35.5</b> | [28.0, 44.5] |
| Syndromes              | Q870-Q879                       | 5.1   | 2.4  | 2.4  | 2.3  | 4.4  | <b>3.3</b>  | [1.3, 6.8]   |

**Table 8 ctd. Anomalies by PCT 2006-2010: rates per 10,000 births****Nottinghamshire County**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 20.9  | 23.1 | 15.8 | 18.4 | 16.6 | <b>18.9</b> | [14.8, 23.8] |
| Eye, ear               | Q100-Q179                       | 0.0   | 0.0  | 1.3  | 0.0  | 0.0  | <b>0.3</b>  | -            |
| Cardiovascular         | Q200-Q269                       | 55.6  | 42.2 | 34.2 | 46.1 | 38.2 | <b>43.1</b> | [36.7, 50.3] |
| Serious Cardiac        | Q20 various (see Table 10)      | 25.0  | 23.1 | 25.0 | 30.3 | 22.9 | <b>25.3</b> | [20.5, 30.9] |
| Urogenital             | Q500-Q649                       | 27.8  | 42.2 | 30.3 | 21.1 | 25.5 | <b>29.3</b> | [24.1, 35.3] |
| Gastrointestinal       | Q350-Q459                       | 25.0  | 24.5 | 14.5 | 17.1 | 22.9 | <b>20.8</b> | [16.4, 25.9] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 58.4  | 76.2 | 64.5 | 68.5 | 35.7 | <b>60.4</b> | [52.8, 68.8] |
| Respiratory            | A300-Q349                       | 7.0   | 6.8  | 2.6  | 3.9  | 0.0  | <b>4.0</b>  | [2.2, 6.6]   |
| Chromosomal            | Q900-Q999                       | 44.5  | 47.6 | 35.5 | 42.1 | 33.1 | <b>40.4</b> | [34.3, 47.4] |
| Syndromes              | Q870-Q879                       | 1.4   | 2.7  | 2.6  | 0.0  | 1.3  | <b>1.6</b>  | [0.6, 3.5]   |

**Rotherham**

| Anomaly Group          | ICD-10                          | Rates |      |      |      |      | TOTAL       |              |
|------------------------|---------------------------------|-------|------|------|------|------|-------------|--------------|
|                        |                                 | 2006  | 2007 | 2008 | 2009 | 2010 | Rate        | 95% C.I.     |
| Central Nervous System | Q000-Q079                       | 36.5  | 37.1 | 15.2 | 28.9 | 30.8 | <b>29.6</b> | [21.8, 39.3] |
| Eye, ear               | Q100-Q179                       | 0.0   | 3.1  | 0.0  | 6.4  | 0.0  | <b>1.9</b>  | -            |
| Cardiovascular         | Q200-Q269                       | 26.6  | 49.5 | 24.4 | 48.2 | 58.5 | <b>41.6</b> | [32.2, 52.8] |
| Serious Cardiac        | Q20 various (see Table 10)      | 19.9  | 15.5 | 12.2 | 35.4 | 33.9 | <b>23.3</b> | [16.4, 32.1] |
| Urogenital             | Q500-Q649                       | 13.3  | 40.2 | 15.2 | 22.5 | 27.7 | <b>23.9</b> | [16.9, 32.8] |
| Gastrointestinal       | Q350-Q459                       | 33.2  | 21.7 | 15.2 | 32.2 | 55.4 | <b>31.5</b> | [23.3, 41.5] |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 29.9  | 55.7 | 54.9 | 41.8 | 36.9 | <b>44.1</b> | [34.4, 55.7] |
| Respiratory            | A300-Q349                       | 6.6   | 3.1  | 6.1  | 9.6  | 12.3 | <b>7.6</b>  | [3.9, 13.2]  |
| Chromosomal            | Q900-Q999                       | 26.6  | 9.3  | 21.3 | 28.9 | 18.5 | <b>20.8</b> | [14.3, 29.2] |
| Syndromes              | Q870-Q879                       | 6.6   | 15.5 | 6.1  | 3.2  | 9.2  | <b>8.2</b>  | [4.4, 14.0]  |

**Sheffield**

| Anomaly Group          | ICD-10                          | Rates |       |       |       |      | TOTAL        |                |
|------------------------|---------------------------------|-------|-------|-------|-------|------|--------------|----------------|
|                        |                                 | 2006  | 2007  | 2008  | 2009  | 2010 | Rate         | 95% C.I.       |
| Central Nervous System | Q000-Q079                       | 29.8  | 33.1  | 32.9  | 23.9  | 25.5 | <b>29.0</b>  | [23.5, 35.5]   |
| Eye, ear               | Q100-Q179                       | 6.3   | 4.5   | 9.0   | 1.5   | 1.5  | <b>4.5</b>   | [2.5, 7.5]     |
| Cardiovascular         | Q200-Q269                       | 161.7 | 201.8 | 146.5 | 109.2 | 49.4 | <b>133.4</b> | [121.3, 146.4] |
| Serious Cardiac        | Q20 various (see Table 10)      | 14.1  | 31.6  | 23.9  | 28.4  | 24.0 | <b>24.5</b>  | [19.5, 30.4]   |
| Urogenital             | Q500-Q649                       | 78.5  | 102.4 | 86.7  | 71.8  | 21.0 | <b>72.0</b>  | [63.2, 81.7]   |
| Gastrointestinal       | Q350-Q459                       | 34.5  | 27.1  | 35.9  | 25.4  | 37.5 | <b>32.1</b>  | [26.3, 38.8]   |
| Musculoskeletal        | Q180-Q189, Q380-Q389, Q650-Q799 | 80.1  | 81.3  | 95.7  | 61.3  | 36.0 | <b>70.8</b>  | [62.0, 80.4]   |
| Respiratory            | A300-Q349                       | 7.8   | 9.0   | 9.0   | 6.0   | 4.5  | <b>7.3</b>   | [4.7, 10.8]    |
| Chromosomal            | Q900-Q999                       | 44.0  | 42.2  | 40.4  | 47.9  | 37.5 | <b>42.4</b>  | [35.6, 50.0]   |
| Syndromes              | Q870-Q879                       | 9.4   | 9.0   | 6.0   | 6.0   | 6.0  | <b>7.3</b>   | [4.7, 10.8]    |

## Anomaly Surveillance

For the great majority of anomalies, trends have remained stable or show a slight decrease between 2006 and 2010, reflecting the time lag necessary for the more recently diagnosed anomalies to reach the Register. **The increasing trend evident in renal dysplasia cases, however, has continued,** and although this remains consistent with other EUROCAT registers, it will be kept under review.

The **rising incidence of gastroschisis** in northern Europe also continues, and will be the subject of an EMSYCAR led EUROCAT research programme from Autumn 2012.

**EUROCAT has recently identified an increasing trend in Congenital Cystic Adenomatoid Malformation,** possibly due to improved antenatal detection. **Upper Limb Reduction anomalies have shown a pan-European decrease** throughout the same time period.

**Analysis of EMSYCAR data shows increasing trends in Transposition of the Great Arteries (TGA) and Posterior Urethral Valves/Prune Belly Syndrome (PUV).** Since TGA is one of the serious cardiac anomalies which had previously been identified as underreported, this increase is appropriate. However, if the rate remains above that of the UK and EUROCAT, then investigations will be conducted. While the EMSYCAR rate of PUV is now exactly the same as the BINOCAR mean, absolute numbers of PUV remain too low for reliable statistical analysis.

**Routine ongoing surveillance of EMSYCAR data by EUROCAT during 2010 highlighted one potential cluster of five cleft lip +/- palate cases,** delivered in a two day period in 2009. This was investigated by the EMSYCAR team according to the EUROCAT protocol; the five cases were validated, but found to be completely independent of one another.

At the request of a PCT, an **investigation was made into a suggested cluster of upper limb reduction defects.** The results of this enquiry were twofold; firstly, whilst the cases had occurred in fairly close spatial proximity, the time scale and differing aetiologies meant that they were unrelated, and secondly, that although two of the three cases were known to EMSYCAR, the procedures for reporting anomalies from the local delivery suite required strengthening, particularly while a relevant computer system awaited an upgrading exercise.

## The NHS Fetal Anomaly Screening Programme and the Antenatal Detection of Congenital Anomalies

In January 2010 the NHS Fetal Anomaly Screening Programme (FASP) published a set of National Standards and Guidance for the 18<sup>+0</sup> to 20<sup>+6</sup> weeks of gestational age Fetal Anomaly Scan in England.

This document provided a set of **eleven conditions**, each capable of being detected antenatally, considered 'important, being fatal, associated with morbidity or requiring immediate postnatal support'. From April 2010, these conditions were to be screened for and audited by ultrasound departments. Target antenatal detection rates vary, but all were set at 50% or greater.

**Table 9: Fetal Anomaly Screening Programme Auditable Conditions**

| Condition                     | Target Detection Rate % |
|-------------------------------|-------------------------|
| Anencephaly                   | 98                      |
| Open Spina Bifida             | 90                      |
| Cleft Lip                     | 75                      |
| Diaphragmatic Hernia          | 60                      |
| Gastroschisis                 | 98                      |
| Exomphalos                    | 80                      |
| Serious Cardiac Anomalies     | 50                      |
| Bilateral Renal Agenesis      | 84                      |
| Lethal Skeletal Dysplasia     | 60                      |
| Edwards Syndrome (Trisomy 18) | 95                      |
| Patau Syndrome (Trisomy 13)   | 95                      |

A number of difficulties which became apparent during this exercise were addressed in the previous EMSYCAR Report for 2005-2009. In this present report, we have continued to define 'lethal skeletal dysplasia' as thanatophoric dysplasia (Q771) and short-rib/Jeune's syndrome (Q772) only, and to employ the EUROCAT definition of 'Serious' cardiac anomalies (see below).

**Table 10: Serious Cardiac Conditions – EUROCAT definitions**

| Condition   | ICD-10     |
|---|------------|
| Common Truncus                                    | Q200       |
| Transposition of the Great Arteries               | Q203       |
| Double Inlet Ventricle/Common Ventricle           | Q204       |
| AVSD  | Q212       |
| Fallot's Tetralogy                                | Q213       |
| Pulmonary Valve Atresia                           | Q220       |
| Tricuspid Atresia/Stenosis                        | Q224       |
| Ebstein's Anomaly                                 | Q225       |
| Hypoplastic Right Heart Syndrome                  | Q226       |
| Aortic Valve Atresia/Stenosis                     | Q230       |
| Hypoplastic Left Heart Syndrome                   | Q234       |
| Coarctation of the Aorta                          | Q251-Q2519 |
| Total Anomalous Pulmonary Venous Drainage (TAPVD) | Q262       |

Difficulties remain with the allocation of anomaly cases to individual hospitals, in particular the more serious anomalies, which are more likely to be referred to a number of different units. Tables are therefore presented by the PCT of mother's residence at delivery. As before, **NO inferences should be drawn concerning the relative performance of individual hospital units or hospital trusts.** The decision to base EMSYCAR's reporting of antenatal diagnoses around the PCT of mother's residence will be reviewed in the light of any future NHS reorganisation.

Since anomalies are rare events and affect a small proportion of live and stillbirths, a very small number of anomalies occur within an individual PCT (or unit) within any one year, and a single pregnancy can have a significant effect on whether or not a specific antenatal detection target is achieved. There may be many different reasons for an apparently 'late' or 'missed' diagnosis, including:

- Mothers who book too late for screening
- Mothers who transfer into an area in the later stages of a pregnancy
- Mothers who decline antenatal screening
- Mothers who decline invasive testing

To address this issue, extra data items have therefore been added to the EMSYCAR Notification Form from the beginning of 2012 to enable more accurate data to be collected. We are most grateful to all the antenatal reporters for supplying this extra information.

Although a pregnancy may be diagnosed antenatally with a potential anomaly, the precise nature of the anomaly may not be confirmed until later in the pregnancy or following delivery. Where this is obvious from the notifications sent to EMSYCAR, a pragmatic view has been taken and the main anomaly under review has been assumed to be antenatally diagnosed.

For example, a case of cleft lip, or AVSD diagnosed antenatally does not always mean that the underlying condition of Trisomy 13 or 21 was also recognised, and some of the chromosomal antenatal diagnosis rates shown here may be slightly inflated as a result. Conversely, an antenatal notification of Trisomy 21 that fails to mention a known AVSD would lead to an apparently lower antenatal diagnosis rate of cardiac anomalies in that PCT/Unit. **It is therefore vital that notifiers report ALL antenatally diagnosed or suspected anomalies to EMSYCAR, particularly where these involve, or might involve, anomalies on the FASP list.**

In 2010, across the whole EMSYCAR region, **70% of all FASP cases were detected antenatally.** The figure for individual PCTs varied between 59% and 81%. This difference is almost certainly related to the higher proportion of 'unknown' cases in some PCTs than others. When reminders are not completed and returned accurately and/or promptly, it can be very difficult for EMSYCAR to establish whether or not a case has been diagnosed antenatally or not. Numbers for individual anomalies within the FASP are too small to analyse by individual years and PCTs, so have been aggregated for the 2006-2010 period in Table 12 (below).

**Table 11: Fetal Anomaly Screening Programme (FASP) Targets for Antenatal Detection of Congenital Anomalies**

|                                  | FASP Target<br>% Antenatal<br>Detection<br>Rate | EMSYCAR Region<br>% Antenatal<br>Detection Rate<br>2006-2010 |
|----------------------------------|---|--|
| Anencephaly                      | 98  | 97.3   |
| Open Spina Bifida                | 90  | 91.1   |
| Cleft Lip                        | 75  | 73.8   |
| Diaphragmatic Hernia             | 60  | 76.4   |
| Gastroschisis                    | 98  | 92.6   |
| Exomphalos                       | 80  | 88.8   |
| Serious Cardiac<br>Anomalies     | 50  | 47.3   |
| Bilateral Renal<br>Agenesis      | 84  | 85.9   |
| Lethal Skeletal<br>Dysplasia*    | 60  | 100  |
| Edwards syndrome -<br>Trisomy 18 | 95  | 86.9   |
| Patau syndrome -<br>Trisomy 13   | 95  | 93.8   |



**Table 12: Antenatal Detection of FASP anomalies by PCT 2006-2010**

|                         | Anencephaly                       | Spina Bifida                      | Serious Cardiac Defect            | Cleft Lip                         | Bilateral Renal Agenesis          | Lethal Skeletal Dysplasia         |
|-------------------------|-----------------------------------|-----------------------------------|-----------------------------------|-----------------------------------|-----------------------------------|-----------------------------------|
| Target detection (%)    | 98                                | 90                                | 50                                | 75                                | 84                                | 60                                |
| PCT                     | rate per 10,000<br>% A/N detected | rate per 10,000<br>% A/N detected | rate per 10,000<br>% A/N detected | rate per 10,000<br>% A/N detected | rate per 10,000<br>% A/N detected | rate per 10,000<br>% A/N detected |
| Barnsley                | 4.26<br>100.0                     | 6.40<br>77.8                      | 20.61<br>65.5                     | 14.93<br>85.7                     | 0.71<br>100.0                     | 0.00<br>-                         |
| Bassetlaw               | 6.56<br>100.0                     | 11.48<br>85.7                     | 14.76<br>55.6                     | 13.12<br>75.0                     | 1.64<br>100.0                     | 1.64<br>100.0                     |
| Derby City              | 1.72<br>66.7                      | 2.87<br>80.0                      | 23.54<br>31.7                     | 12.63<br>77.3                     | 0.00<br>-                         | 0.57<br>100.0                     |
| Derbyshire County       | 4.09<br>93.8                      | 6.90<br>85.2                      | 19.42<br>43.4                     | 5.62<br>59.1                      | 1.02<br>100.0                     | 0.00<br>-                         |
| Doncaster               | 7.97<br>100.0                     | 6.91<br>84.6                      | 14.87<br>50.0                     | 19.66<br>67.6                     | 2.66<br>80.0                      | 0.53<br>100.0                     |
| Leicester City          | 8.18<br>100.0                     | 8.18<br>95.2                      | 28.06<br>37.5                     | 10.91<br>67.9                     | 3.12<br>87.5                      | 1.56<br>100.0                     |
| Leicestershire County   | 3.59<br>100.0                     | 4.70<br>94.1                      | 21.82<br>41.8                     | 8.84<br>81.3                      | 0.83<br>33.3                      | 0.28<br>100.0                     |
| Lincolnshire            | 4.29<br>100.0                     | 5.90<br>95.5                      | 19.30<br>34.7                     | 8.04<br>73.3                      | 0.80<br>100.0                     | 1.07<br>100.0                     |
| North Lincolnshire      | 4.29<br>100.0                     | 5.36<br>80.0                      | 21.44<br>40.0                     | 9.65<br>100.0                     | 2.14<br>100.0                     | 3.22<br>100.0                     |
| North East Lincolnshire | 5.02<br>100.0                     | 8.04<br>100.0                     | 18.08<br>27.8                     | 17.08<br>52.9                     | 3.01<br>100.0                     | 0.00<br>-                         |
| Northamptonshire        | 5.51<br>100.0                     | 5.07<br>95.7                      | 21.61<br>54.1                     | 6.17<br>85.7                      | 1.54<br>85.7                      | 0.88<br>100.0                     |
| Nottingham City         | 3.32<br>71.4                      | 3.79<br>87.5                      | 25.11<br>67.9                     | 8.53<br>66.7                      | 0.95<br>100.0                     | 0.47<br>100.0                     |
| Nottinghamshire County  | 3.73<br>92.9                      | 6.12<br>95.7                      | 25.28<br>65.3                     | 7.98<br>66.7                      | 0.53<br>100.0                     | 0.00<br>-                         |
| Rotherham               | 6.93<br>100.0                     | 11.96<br>89.5                     | 23.30<br>32.4                     | 5.67<br>66.7                      | 2.52<br>100.0                     | 0.00<br>-                         |
| Sheffield               | 6.05<br>100.0                     | 4.84<br>100.0                     | 24.50<br>46.9                     | 9.98<br>84.8                      | 2.72<br>100.0                     | 1.21<br>100.0                     |
| <b>EMSYCAR Total</b>    | <b>4.91</b><br>97.2               | <b>6.08</b><br>91.5               | <b>22.02</b><br>47.4              | <b>9.37</b><br>73.8               | <b>1.47</b><br>90.7               | <b>0.65</b><br>100.0              |

Shaded boxes indicate rates of antenatal detection between 2006-2010 below FASP target for 2011.

**Table 12 ctd. Antenatal Detection of FASP Anomalies by PCT 2006-2010**

|                         | Diaphragmatic<br>Hernia                     | Exomphalos                                  | Gastroschisis                               | Trisomy<br>18                               | Trisomy<br>13                               |
|-------------------------|---|---|---|---|---|
| Target detection (%)    | 60  | 80  | 98  | 95  | 95  |
| PCT                     | rate per<br>10,000<br><br>% A/N<br>detected | rate per<br>10,000<br><br>% A/N<br>detected | rate per<br>10,000<br><br>% A/N<br>detected | rate per<br>10,000<br><br>% A/N<br>detected | rate per<br>10,000<br><br>% A/N<br>detected |
| Barnsley                | 3.55<br>80.0                                | 3.55<br>100.0                               | 7.82<br>100.0                               | 2.84<br>100.0                               | 1.42<br>100.0                               |
| Bassetlaw               | 4.92<br>33.3                                | 4.92<br>100.0                               | 8.20<br>100.0                               | 4.92<br>66.7                                | 4.92<br>100.0                               |
| Derby City              | 1.72<br>66.7                                | 5.17<br>88.9                                | 4.02<br>100.0                               | 4.59<br>62.5                                | 2.30<br>100.0                               |
| Derbyshire County       | 2.56<br>80.0                                | 2.56<br>80.0                                | 6.39<br>92.0                                | 2.56<br>90.0                                | 2.56<br>100.0                               |
| Doncaster               | 3.72<br>100.0                               | 4.25<br>87.5                                | 9.56<br>94.4                                | 5.84<br>100.0                               | 2.66<br>100.0                               |
| Leicester City          | 3.90<br>50.0                                | 5.46<br>85.7                                | 6.63<br>94.1                                | 6.24<br>87.5                                | 2.34<br>100.0                               |
| Leicestershire County   | 1.38<br>40.0                                | 4.14<br>93.3                                | 3.59<br>76.9                                | 7.74<br>89.3                                | 3.87<br>85.7                                |
| Lincolnshire            | 2.41<br>88.9                                | 3.22<br>91.7                                | 5.36<br>90.0                                | 4.83<br>72.2                                | 3.22<br>100.0                               |
| North Lincolnshire      | 4.29<br>100.0                               | 1.07<br>100.0                               | 5.36<br>100.0                               | 1.07<br>100.0                               | 0.00<br>-                                   |
| North East Lincolnshire | 2.01<br>100.0                               | 2.01<br>100.0                               | 8.04<br>100.0                               | 1.00<br>100.0                               | 1.00<br>100.0                               |
| Northamptonshire        | 3.75<br>82.4                                | 5.95<br>81.5                                | 3.09<br>92.9                                | 5.07<br>87.0                                | 2.20<br>90.0                                |
| Nottingham City         | 3.79<br>87.5                                | 3.32<br>100.0                               | 5.69<br>91.7                                | 9.00<br>94.7                                | 3.32<br>100.0                               |
| Nottinghamshire County  | 5.59<br>81.0                                | 6.39<br>83.3                                | 3.46<br>84.6                                | 5.06<br>94.7                                | 2.39<br>88.9                                |
| Rotherham               | 2.52<br>75.0                                | 3.78<br>100.0                               | 10.07<br>93.8                               | 2.52<br>50.0                                | 1.89<br>100.0                               |
| Sheffield               | 4.54<br>66.7                                | 5.45<br>94.4                                | 6.05<br>95.0                                | 10.29<br>88.2                               | 3.33<br>81.8                                |
| <b>EMSYCAR Total</b>    | <b>3.35</b><br><b>76.4</b>                  | <b>4.39</b><br><b>88.8</b>                  | <b>5.56</b><br><b>92.6</b>                  | <b>5.42</b><br><b>86.9</b>                  | <b>2.64</b><br><b>93.8</b>                  |

Shaded boxes indicate rates of antenatal detection between 2006-2010 below FASP target for 2011.

## Ongoing EMSYCAR Activities

**Audit and research activities** have continued using Register data. EMSYCAR has joined several new European-wide research initiatives, including one investigating the epidemiology of Hirschsprungs' Disease, and a number of collaborative papers (see below) have been published using EMSYCAR data with EMSYCAR staff as collaborating authors.

**The EMSYCAR team** will also, from Autumn 2012, be leading the research for a section of EUROCAT work package 7, investigating the changing epidemiology of gastroschisis, an anomaly which is known to have a higher birth prevalence in the UK and Northern Europe than elsewhere.

**The EMSYCAR team have published a paper in the BMJ** entitled 'Socioeconomic inequalities in outcome of pregnancy and neonatal mortality associated with congenital anomalies: population based study'. This used data from the Register for cases with one of nine congenital anomalies with poor prognostic outcome between 1998-2007 to demonstrate that socioeconomic variation in decisions regarding termination of pregnancy after antenatal detection of a serious anomaly has resulted in widening socioeconomic inequality in live born infants and subsequent neonatal mortality.

**An application for funding** to the College of Medicine and Biological Sciences at Leicester University **for a PhD student** to pursue this subject in more depth was subsequently successful. The post was advertised, and an appointment made from the beginning of October 2012.

**EMSYCAR data for 2009-2011 has been used by the Yorkshire Cardiac Network** to help evaluate the outcomes of its training programme for sonographers, by comparing rates of antenatal diagnosis for a range of cardiac anomalies both before and after the training was implemented.

**EMSYCAR continues to contribute to a number of ongoing UK projects**, and recent publications have included the monitoring and prenatal detection of structural fetal congenital anomalies in England and Wales, an audit of congenital diaphragmatic hernia, and the results of a collaborative investigation into the antenatal diagnosis of schizencephaly. EMSYCAR has also contributed to a **BINOCAR audit of Down syndrome** cases between 2003 and 2006 and continues to participate with an investigation into the **rising incidence of gastroschisis**.

**EMSYCAR-led research projects continue** with an audit of all cases of Turner syndrome reported to the Register, together with an associated review of all the reported cases of cystic hygroma. Initially conceived as an update of a paper published by the Leicester team in 2005, this has already resulted in the award of first prize for a poster at the EUROCAT Scientific Conference in Antwerp, 2011 (see below). This submission was rewarded as the contribution most likely to make a positive impact on patient management and clinical practice.

A **new audit**, initiated by Fetal Medicine specialists in Leicester, is being undertaken **to establish the risk of congenital anomalies in discordant twin pregnancies**.

**Recent EMSYCAR data has been used by the NHS Fetal Anomaly Screening Programme** in a review of antenatal diagnosis rates for selected congenital anomalies. Along with West Midlands CAR and the North-Eastern Register, NORCAS, EMSYCAR has co-operated in the production of a

detailed report to FASP, outlining “A methodology to support the monitoring of the Fetal Anomaly Screening programme and prenatal detection of anomalies in England”, which was submitted in May 2012.

**EMSYCAR is now providing expertise on anomaly coding and definition** to a group including fetal medicine specialists, radiologists, paediatricians, pathologists and cardiologists, tasked with producing a workable definition of ‘serious cardiac anomalies’ capable of being identified antenatally, and therefore monitored, by the FASP.

**EMSYCAR data**, with other BINOCAR Registers, has been utilised throughout 2011 and 2012 by the National Perinatal Epidemiology Unit in Oxford, **to validate data obtained from BAPS-CASS** (the British Association of Paediatric Surgeons Congenital Anomalies Surveillance System). This has attempted to evaluate the success of the BAPS-CASS data collection methods.

Regular data matching and cleaning is undertaken in conjunction with the North Trent Clinical Genetics Service, the National Down Syndrome Cytogenetic Register, together with local ultrasound, antenatal and fetal medicine units, mainly with a view to establishing antenatal detection rates. Training sessions have continued to be provided at the request of local clinicians. **Data requests are regularly received** from local clinicians and antenatal screening co-ordinator, and have included requests for

- details of rates of urogenital anomalies within a PCT
- cases of tracheo-oesophageal fistula occurring within a unit
- antenatal detection of cardiac anomalies within a unit
- cases reported with echogenic bowel within a unit
- baseline data for a summary report into the effects of Schmallenburg Virus
- EMSYCAR cases of Fraser syndrome
- socio-economic variation in anomaly rates within a PCT
- variation in rates of antenatal detection of TGA between units

Professor Elizabeth Draper continues in her role as **Chair of the British Isles Network of Congenital Anomaly Registers (BINOCAR)**. Dr Judith Budd continues as a member of the **EUROCAT Coding Issues Group and BINOCAR Coding Committee**. EMSYCAR was represented at the EUROCAT Annual Register Leaders’ Meetings in Antwerp and Budapest in 2011 and 2012.

## **Papers using EMSYCAR data and/or by EMSYCAR staff published since last Annual Report:**

1. Pedersen R, Calzolari E, Husby S et al. "Oesophageal atresia: prevalence, prenatal diagnosis and associated anomalies in 23 European regions", Arch.Dis.Child. (published online 13<sup>th</sup> January 2012) doi: 10.1136/archdischild-2011-300597
2. Best K, Tennant P., Addor MC. et al. "Epidemiology of small intestinal atresia in Europe: a register based study", Arch.Dis.Child.Fetal.Neonatal.Ed (2012) doi: 10.1136/archdischild-2011-300631.
3. Howe D, Rankin J & Draper E. "Schizencephaly prevalence, prenatal diagnosis and clues to etiology: a register based study", Ultrasound.Obstet.Gynecol (2012) 39: 75-82
4. Loane M, Morris J, Addor MC et al. "Twenty year trends in the prevalence of Down syndrome and other trisomies in Europe: impact of maternal age and prenatal screening", Eur.J.Hum.Genet. (2012) June 20<sup>th</sup> doi: 10.1038/ejhg.2012.94
5. Navti O, Kinning E, Vasudevan P et al. "Review of perinatal management of arthrogyrosis at large UK teaching hospital serving a multi-ethnic population", Prenatal Diagnosis (2010) 30: 49-56.
6. Dolk H, Loane M, Garne E and a EUROCAT Working Group. "Congenital Heart Defects in Europe: Prevalence and Perinatal Mortality, 2000-2005", Circulation (2011) 123: 841-849
7. Wellesley D, Dolk H, Boyd P. et al. "Rare chromosome abnormalities, prevalence and prenatal diagnosis rates from population-based congenital anomaly registers in Europe", Eur.J.Hum.Genet. (2012) 20, 521-526 doi. 10.1038/ejhg.2011.246
8. Smith L, Budd J, Field D & Draper E. "Socio-economic inequalities in outcome of pregnancy and neonatal mortality associated with congenital anomalies: population based study", BMJ (2011) 343:d4306 doi: 10.1136/bmj.d4306

## **EMSYCAR Poster Presentations since last Annual Report:**

1. Budd J, Craft E & Draper E. "Outcomes following prenatal diagnosis of Cystic Hygroma" – awarded First Prize at the 11<sup>th</sup> European Symposium on Congenital Anomalies in Antwerp, June 2011, as the contribution most likely to impact positively on patient care and management.
2. Smith L, Budd J., Field D & Draper E. "Socioeconomic variation in termination of pregnancy, live birth and neonatal mortality associated with congenital anomalies" – presented to the Annual Conference of the British Maternal and Fetal Medicine Society, Harrogate, June 2011
3. As above, presented to the Society of Perinatal Epidemiological Research (SPER), and the Society for Epidemiologic Research (SER) in Montreal, June 2011.
4. Craft E, Budd J & Draper E. "Outcomes following the diagnosis of Turner Syndrome: EMSYCAR". Presented to the Annual Conference of the British Society for Human Genetics, York, 2011.

5. Budd J, Abou El Senoun G, Berry L, Mousa H & Draper E. "Antenatal Detection of Facial Clefts: is the FASP target achievable?" Presented to the Annual Conference of the British Maternal and Fetal Medicine Society, Glasgow, 2012.
6. Yadav K, Vasudev D, Budd J & Akuma A. "Congenital Cystic Adenomatoid Malformation: is it always bad news?" Presented to the Annual Conference of the British Maternal and Fetal Medicine Society, Glasgow, 2012.
7. Berry L, Budd J, Perry P & Draper E. "The Importance of Congenital Anomaly Surveillance: EMSYCAR". Presented at the 1<sup>st</sup> Annual Festival of Public Health, Manchester, 2012.

### **EMSYCAR Conference/Meeting Presentations since last Annual Report:**

1. Budd J., Smith L, Field D & Draper E. "Socioeconomic Inequalities in the risk of congenital anomalies and pregnancy outcome". Presented to the 14th International Medical Geography Symposium, Durham, July 2011.
2. As above, also presented at the 1<sup>st</sup> Yorkshire and Humber CAR Education Day, Leeds, December 2011.
3. Draper E . "The prevalence of limb reduction deformities: EMSYCAR, BINOCAR & EUROCAT". Presented at the East Midlands Fetal Medicine Meeting, Nottingham, March 2012.