Third report of the

Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire (CAROBB)

Births in 2005-2010

Births within Oxfordshire 1991-2010

March 2012

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The report can be accessed at website: www.npeu.ox.ac.uk/carobb/

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The CAROBB Team

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Confidentiality and policy on non-disclosure of small numbers

As a member of BINOCAR (British Isles Network of Congenital Anomaly Registers), CAROBB has the approval of the Trent MREC and the National Information Governance Board (NIGB) to collect identifiable information without explicit consent from individuals registered. See documentation in Appendix 5.

We have followed the BINOCAR policy concerning the disclosure of small numbers (www.binocar.org/methods/dataconfidentiality).

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Part 1 - Introduction and Summary

Introduction

This report provides data on prenatally suspected and postnatally confirmed congenital anomalies from cases notified to the Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire (CAROBB) for births occurring in the six years from 2005 – 2010. It also provides 20 years of data (1991 to 2010) from within Oxfordshire (Appendix 1).

In April 2003 the Department of Health awarded funding for the expansion and development of the Oxford Congenital Anomaly Register (OXCAR), for research purposes. A new population-based register, covering the three counties which make up Thames Valley was formed, called the Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire (CAROBB). CAROBB is based at the National Perinatal Epidemiology Unit (NPEU), University of Oxford. This is the third full report from CAROBB and provides population based information on congenital anomalies affecting births between 2005 and 2010 to mothers resident in the three counties.

The main change that affects the register since the last report in 2009 is that the National Congenital Anomaly System (NCAS), funded by the Department of Health (DH) ceased to function in 2010. NCAS, set up in 1964 in response to the Thalidomide disaster, was responsible for surveillance of congenital anomalies in England and Wales. The British Isles Network of Congenital Anomaly Registers (BINOCAR www.binocar.org) has now been funded by the DH to provide surveillance in England and Wales in the areas covered by BINOCAR registries. CAROBB is now the only source of data for surveillance in the three counties. Data for surveillance purposes and research are also sent to the European Congenital Anomaly Surveillance System (EUROCAT, www.eurocat.ulster.ac.uk).

Since the last report we are pleased to have appointed Yvonne Kenworthy as research midwife to CAROBB. Yvonne is in regular contact with all the clinical areas providing data and is exploring with the clinical staff ways of improving our ascertainment. Please contact us if you have any suggestions regarding improvement of ascertainment or if you are interested in using the data for audit or research purposes. For details of projects that CAROBB is involved with, publications to which CAROBB has contributed, ethical approval for CAROBB and how to request data, please see Appendices 3 - 5.

The principal objectives of CAROBB

are to:

- Provide data for research on the aetiology and natural history of congenital anomalies to enable better advice based on accurate information to be given to parents and prospective parents.
- Enable the evaluation and monitoring of new invasive and non-invasive prenatal diagnostic tests and screening programmes.
- Provide data for health care policies and planning.
- Provide data to investigate clusters of abnormalities and putative teratogens by the monitoring of rates over time and of population trends such as maternal age, ethnicity, and health inequalities.
- Improve ascertainment to BINOCAR and to EUROCAT.

The population studied for this report

- This report has information on congenital anomalies suspected and/or confirmed in fetuses / babies born to mothers resident in the three counties of Thames Valley (Oxfordshire, Berkshire and Buckinghamshire) at the time of the birth of the baby, the geographical area of CAROBB.

- Data are provided on cases notified to CAROBB by December 2011 and with a date of birth/delivery 2005-2010 inclusive. For this report a 'case' is a birth with a suspected and / or confirmed congenital anomaly notified to CAROBB. The term 'birth' (unless otherwise stated) is used to cover all pregnancies (from 10 weeks gestation) ending in live birth, stillbirth, miscarriage/intrauterine death and termination of pregnancy for fetal anomaly (TOPFA).

- Denominator data are provided by the Office for National Statistics and include only live births and stillbirths of 24 weeks gestation or more. There were 178,152 total births in Thames Valley between 2005 and 2010.

- The proportion of births with congenital anomalies are given as a percentage of total births or as a rate per 1,000 total births.

The report gives data on anomalies, their rate and, where appropriate, their prenatal detection, in Oxfordshire, Berkshire and Buckinghamshire (Thames Valley). Information on cases for the hospital at which the mother booked for delivery can be provided and presented at the individual hospitals.

Definition and coding of congenital anomalies

The definition of congenital anomaly, used by CAROBB is 'a structural or functional anomaly, presumed to be of prenatal origin'. All anomalies present at birth or diagnosed after birth are recorded. Prenatally suspected anomalies including ultrasound 'soft markers' (normal variants) are also recorded including those occurring in cases subsequently confirmed to be structurally normal babies. In line with other British and European registers each anomaly is coded using the ICD10 classification with the BPA extensions where appropriate.

Summary

- From January 2005 to December 2010 there were 3753 births with a confirmed congenital anomaly (2.1% of all births), to mothers resident in Thames Valley, notified to CAROBB.

- In 62% of these births there was some prenatal suspicion of congenital anomaly.

- One thousand and seventy two births (29% of all births notified with a congenital anomaly) were terminations of pregnancy for fetal anomaly.

- More male than female births were affected by a congenital anomaly, M:F = 1.3:1

- There were 512 births with Down's syndrome; 305 (60%) were prenatally diagnosed. A high risk first trimester screening test result was the most common reason for prenatal diagnosis. Taking into account those cases with a positive Down's syndrome screening test or suspicion on ultrasound scan but where no karyotyping was performed, the potential prenatal detection rate was 72%.

- Research using CAROBB (and previously OXCAR) data is reported in Appendices 3 and 4.

- We recognise that there is some under ascertainment of postnatally diagnosed anomalies to CAROBB, particularly cardiac anomalies diagnosed after the mother has been discharged from the maternity hospital and those not requiring surgery under the age of one year. Births to mothers resident in Thames Valley but delivering outside the CAROBB area (e.g. in London) may not at present be notified.

Table 1 Prenatal detection of specific congenital anomalies in Thames Valley, 2005 – 2010

Anomaly	Test performed	Number of pregnancies notified with prenatal suspicion of anomaly[1]	Number of cases notified with anomaly confirmed at birth	Prevalence per 1,000 total births	Prenatal detection rate[2]
Isolated neural tube defects	Ultrasound Scanning +/- MS AFP[3]	157	165	0.9	95%
Isolated cardiac anomaly	Ultrasound scanning	177	562	3.2[4]	31%
Isolated cleft lip +/- palate	Ultrasound scanning	88	126	0.7	70%
Down's Syndrome	Karyotyping, screening tests, ultrasound scanning	305[2]	512	2.6	60%
Isolated diaphragmatic hernia	Ultrasound scanning	24	35	0.2	69%
Isolated exomphalos	Ultrasound scanning +/- MS AFP	26	29	0.2	90%
Isolated gastroschisis	Ultrasound scanning +/- MS AFP	55	55	0.3	100%

[1]Not including false positive diagnoses
[2]Only includes those karyotyped prenatally – not those with high risk screening result or suspicion on scan who were not karyotyped
[3]MS AFP Maternal Serum Alpha Feto Protein screening- now not routinely performed
[4]Low prevalence because of low ascertainment of cases diagnosed after birth.

Part 2 - Routine statistics, area covered by CAROBB and outcome of pregnancies

Population and area covered

There were over two million people resident in Thames Valley between 2005 and 2010, with Berkshire having the largest and Oxfordshire the smallest population. The numbers in Tables 2 and 3 are supplied by the Office for National Statistics.

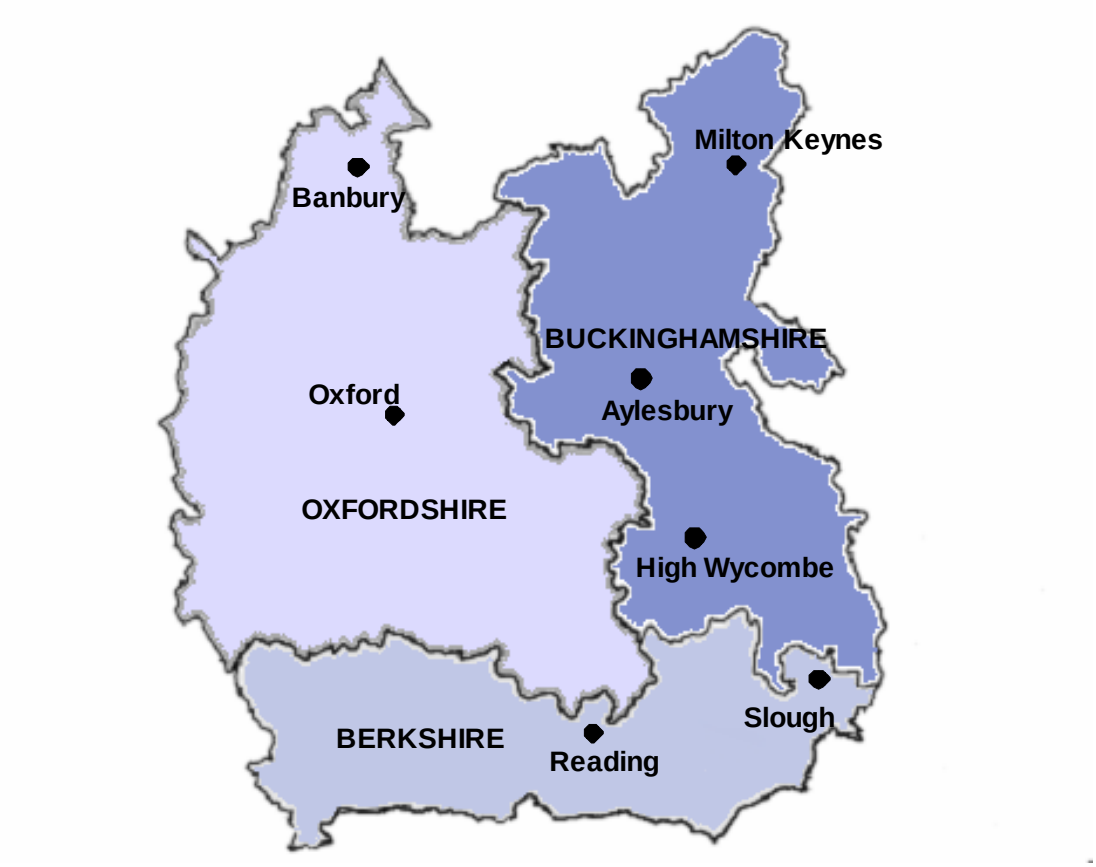
Table 2 Total population covered – mid year estimates by county and year of birth

	Oxfordshire	Berkshire	Buckinghamshire	Thames Valley
2005	627,500	808,800	704,700	**2,141,000**
2006	629,600	817,000	710,100	**2,156,700**
2007	632,300	828,800	717,600	**2,178.700**
2008	635,500	841,800	724,400	**2,201,700**
2009	640,300	854,000	731,400	**2,225,700**
2010	648,700	865,100	739,600	**2,253,400**

Table 3 Total births (live and stillbirths), by county and year of birth

	Oxfordshire	Berkshire	Buckinghamshire	Thames Valley
2005	7616	10920	8762	**27298**
2006	8028	11391	9276	**28695**
2007	8184	12130	9402	**29716**
2008	8347	12490	9893	**30730**
2009	8175	12443	9774	**30392**
2010	8485	12770	10066	**31321**
Total	**48835**	**72144**	**57173**	**178152**

Figure 1 **Map of the CAROBB area, Oxfordshire, Berkshire and Buckinghamshire, forming Thames Valley.**



Total births with congenital anomalies, pre and postnatal diagnosis

Table 4 **Number of cases (% of all births) with congenital anomaly*, by year of birth**

	Oxfordshire n (%)	Berkshire n (%)	Buckinghamshire n (%)	Thames Valley n (%)
2005	174 (2.3%)	164 (1.5%)	173 (2.0%)	**511 (1.9%)**
2006	216 (2.7%)	194 (1.7%)	186 (2.0%)	**596 (2.1%)**
2007	228 (2.8%)	191 (1.6%)	188 (2.0%)	**607 (2.0%)**
2008	247 (3.0%)	205 (1.6%)	190 (1.9%)	**642 (2.1%)**
2009	294 (3.6%)	197 (1.6%)	223 (2.3%)	**714 (2.3%)**
2010	272 (3.2%)	192 (1.5%)	219 (2.2%)	**683 (2.2%)**
Total	**1431 (2.9%)**	**1143 (1.6%)**	**1179 (2.1%)**	**3753 (2.1%)**

*including termination of pregnancy for fetal anomaly

There appears to be a lower rate of congenital anomalies in Berkshire. This almost certainly does not reflect a true reduction in prevalence but is probably due to lower ascertainment, partly because more babies with congenital anomalies born to mothers resident in Berkshire are delivered in London (i.e. outside the Thames Valley area) and although eligible to be notified to CAROBB it is likely that this does not occur for all cases. The rate in Oxfordshire appears higher and this is probably due to the fact that there are well established practices in place for ascertaining cases because a congenital anomaly register (OXCAR) was established in 1991, whereas in Berkshire and Buckinghamshire these are still being set up.

Table 5 illustrates the number and percentage of cases prenatally and postnatally diagnosed. The percentage of cases with a prenatal suspicion of anomaly which were apparently normal at birth is falling. Most of these cases in the early years were associated with ultrasound 'soft markers' (normal variants) such as choroid plexus cysts and the fall probably represents changes in practice, following local protocols and recommendations from the Fetal Anomaly Screening Programme (http://fetalanomaly.screening.nhs.uk/programmestatements) for the reporting of these normal variants.

Table 5 Total births and notifications; number prenatally suspected with and without congenital anomaly at birth and total births with anomalies, by year of birth

Year	2005	2006	2007	2008	2009	2010	Total
Total births	**27298**	**28695**	**29716**	**30730**	**30392**	**31321**	**178152**
Total cases notified to CAROBB*	648	782	760	771	834	771	4566
Number of cases notified prenatally (including normal variants) (% of total notified)	475 (73.3%)	591 (75.6%)	514 (67.6%)	509 (66.0%)	546 (65.5%)	489 (63.3%)	3124 (68.4%)
Number of cases notified prenatally with anomaly confirmed at birth (% of total cases with anomaly)	338 (66%)	405 (68%)	361 (59%)	382 (60%)	432 (61%)	401 (59%)	2319 (62%)
Number of cases notified prenatally & considered normal at birth (% of total notified prenatally)	126 (27%)	172 (29%)	143 (28%)	117 (23%)	107 (20%)	83 (17%)	748 (24%)
Total cases with anomaly at birth, miscarriage or TOPFA (excludes those notified prenatally and lost to follow up) (% of total births)	**511 (1.9%)**	**596 (2.1%)**	**607 (2.0%)**	**642 (2.1%)**	**714 (2.3%)**	**683 (2.2%)**	**3753 (2.1%)**

*Including prenatally suspected cases without anomaly present at birth.

The percentage of births with a congenital anomaly (2.1%) in Table 5 differs from that using the data transferred to EUROCAT (2.0%, see Table 7) because some cases are excluded from analysis by EUROCAT (e.g. those cases resulting in miscarriages before 20 weeks gestation).

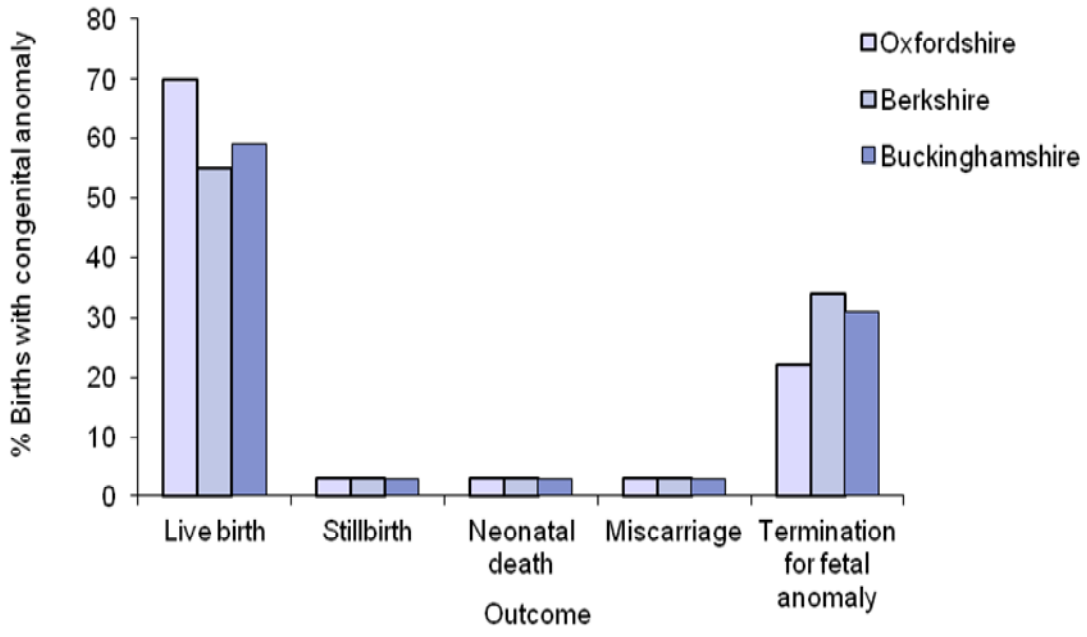
Outcome of pregnancy

Table 6 Outcome of pregnancy of cases notified with congenital anomaly confirmed at birth from 2005 to 2010, by county (n = 3753)^

	Oxfordshire n (%)	Berkshire n (%)	Buckinghamshire n (%)	Thames Valley n (%)
Live birth	1006 (70%)	629 (55%)	691 (59%)	2326 (62%)
Neonatal death	36 (3%)	39 (3%)	40 (3%)	115 (3%)
Stillbirth	33 (3%)	31 (3%)	36 (3%)	100 (3%)
Miscarriage	34 (3%)	37 (3%)	41 (3%)	112 (3%)
Termination for fetal anomaly	313 (22%)	393 (34%)	366 (31%)	1072 (29%)
Not known*	9 (1%)	14 (1%)	5 (<0.5%)	28 (1%)
Total notified	1431	1143	1179	3753

* pregnancies where the diagnosis was known but the pregnancy outcome was not known
^percentages may not add up to 100% because of rounding

Figure 2 Outcome of pregnancy (percentage of live births, stillbirths, neonatal deaths, miscarriages or terminations of pregnancy) with congenital anomaly, 2005-2010, by county, n = 3753

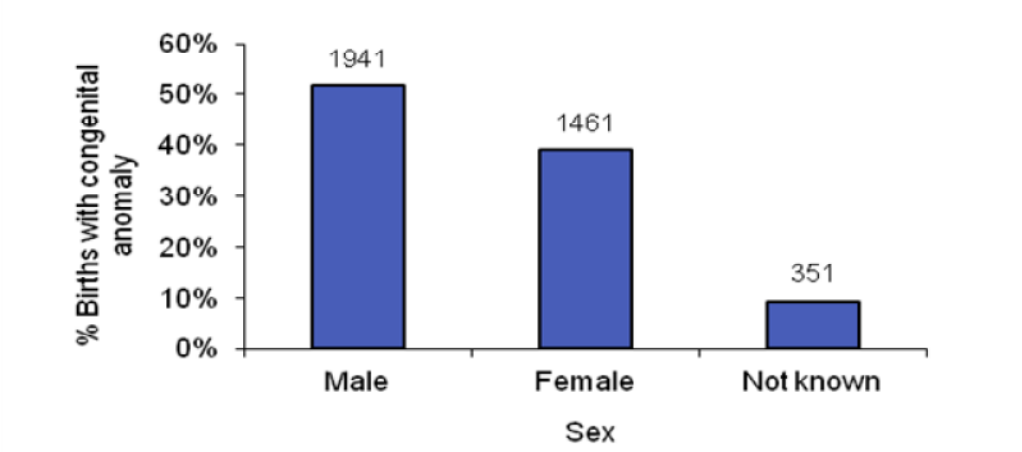


The termination of pregnancy for fetal abnormality rate appears to be lower in Oxfordshire. This is most likely to reflect the improved ascertainment of congenital anomalies diagnosed after birth in Oxfordshire where the register has been collecting data for more than 20 years.

Sex ratio of births with congenital anomalies

Figure 3 Percentage and number of male and female births with congenital anomaly

Sex ratio of cases with anomaly at birth M:F 1.3:1



The sex ratio for births with a congenital anomaly in the CAROBB area, in 2005-2008 is 1.3;1, the same as that for all the other BINOCAR registries in 2009. The background rate for all births in England and Wales in the same time period is 1.13:1.0 (data provided by the Office for National Statistics).

Termination of pregnancy for fetal anomaly (TOPFA), 2005 - 2010

Figure 4a shows the percentage and number of cases resulting in TOPFA by type of anomaly. Chromosome anomalies accounted for 51% of cases, isolated structural anomalies for 34%, single gene defects for 6%, and 7% were non-chromosomal multiple structural anomalies. Of the chromosome anomalies 52% had Down's syndrome (Figure 4b). Neural Tube defects were the most common isolated structural defect resulting in TOPFA (Figure 4c). Ninety one percent of TOPFAs were performed before 25 weeks of gestation (Figure 4d)

Figure 4a Percentage and number of cases resulting in TOPFA by type of anomaly, n = 1072

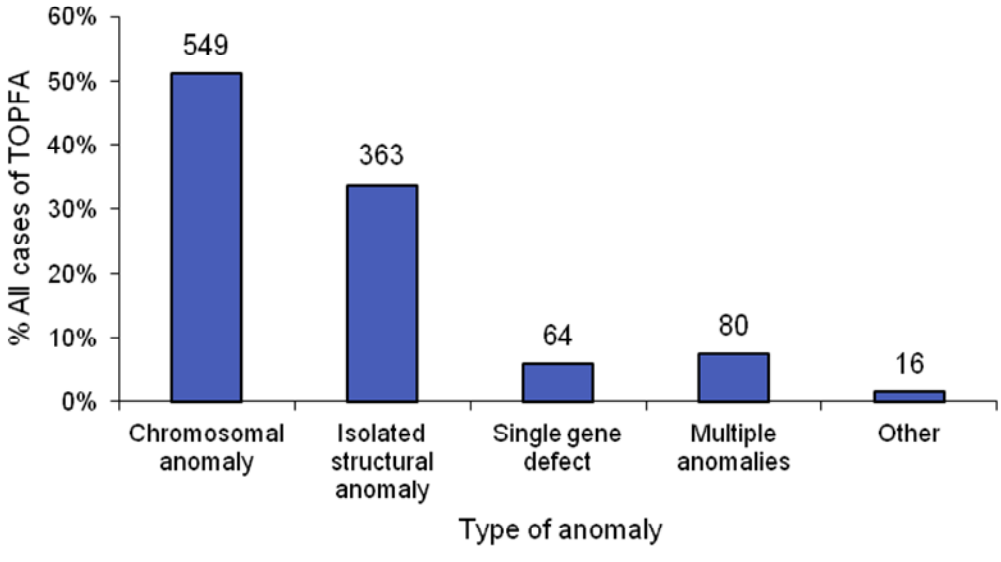


Figure 4b: **TOPFA, chromosome anomalies by type, n = 549**

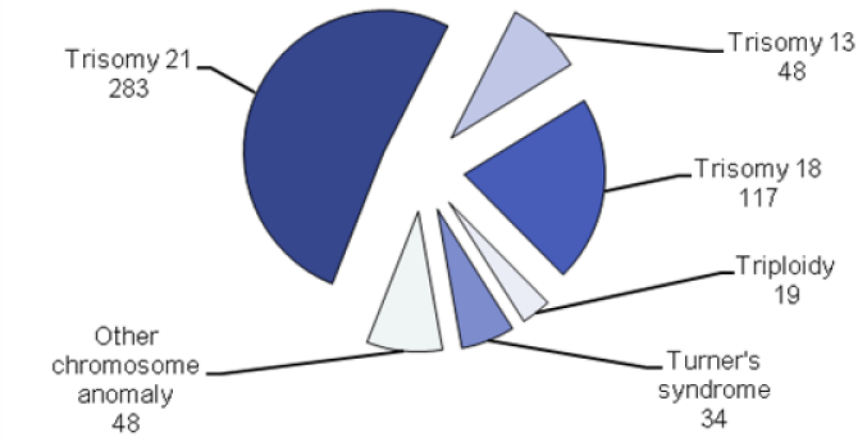


Figure 4c: **TOPFA, isolated anomalies by type, n = 363**

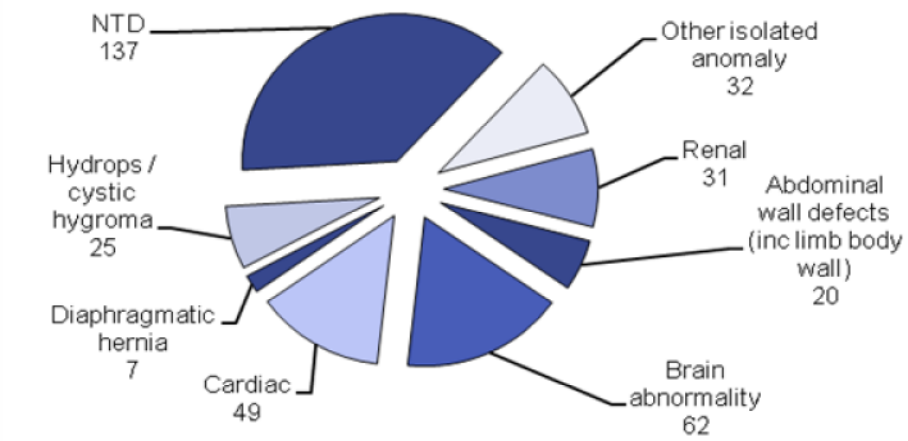
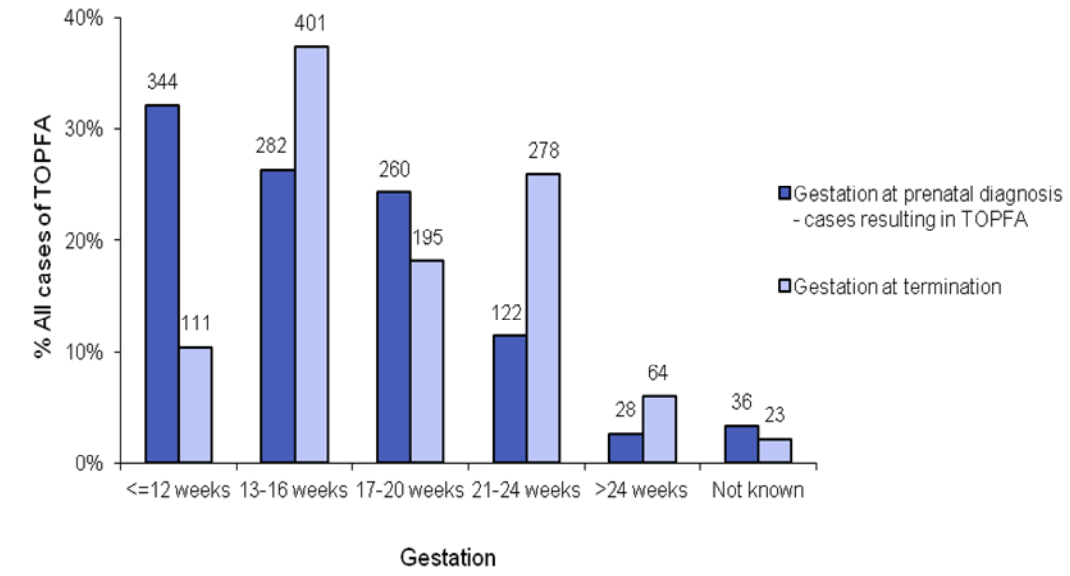


Figure 4d **Percentage and number of cases resulting in termination of pregnancy for fetal anomaly (TOPFA), by gestation period at prenatal diagnosis and at termination,**
n = 1072



Part 3 - Rates of congenital anomalies

Table 7 Table of cases and anomalies and rate per 1,000 births using data from CAROBB held by EUROCAT, year of birth 2005 - 2010 (Total births: 178,152)

Please note: *The reason for the lower the rate of births with congenital anomalies than that shown in Tables 4-6 is that not all births notified to CAROBB are transmitted to EUROCAT e.g. miscarriages of less than 20 weeks of gestation.
^Includes cases where a diagnosis was made but the outcome of pregnancy is not known.

Diagnostic Category	ICD 10 code	Live births, stillbirths and fetal deaths >=20weeks (n)	Termination of pregnancy (n)	Including chromosomal anomalies Rate per 1,000 births		Excluding chromosomal anomalies Rate per 1,000 births	
				Live births, stillbirths, fetal deaths and termination of pregnancy^ (n)	Live births, stillbirths, fetal deaths and termination of pregnancy (rate)	Live births, stillbirths, fetal deaths and termination of pregnancy (n)	Live births, stillbirths, fetal deaths and termination of pregnancy (rate)
All births with congenital anomalies		**2506**	**1028**	**3534**	**19.8**	**2664**	**15.0**
The list below is a list of all anomalies, not individual births. Some births will have more than one anomaly present. An anomaly listed as resulting in termination of pregnancy may be part of a multiple anomaly case.							
Nervous system anomalies	Q00 – Q07	**143**	**275**	**418**	**2.3**	**377**	**2.1**
Neural Tube Defects		33	163	196	1.1	184	1.0
Anencephalus, encephalocoele and similar	Q00 – Q01	11	96	107	0.6	102	0.6
Spina Bifida	Q05	22	67	89	0.5	82	0.5
Hydrocephaly	Q03	49	55	104	0.6	96	0.5
Congenital heart anomalies	Q20 - Q26	**686**	**121**	**807**	**4.5**	**683**	**3.8**
Severe CHD	Q200, Q203, Q204, Q212, Q213, Q225, Q226, Q224, Q220, Q230, Q234, Q251, Q262	254	65	319	1.8	270	1.5
Respiratory anomalies	Q30 – Q34	**76**	**12**	**88**	**0.5**	**83**	**0.5**

Oro-facial clefts	Q35 - Q37	**244**	**29**	**273**	**1.5**	**252**	**1.4**
Digestive system anomalies	Q38 – Q39, Q402, Q408, Q409, Q41 – Q45	**167**	**29**	**196**	**1.1**	**171**	**1.0**
Oesophageal atresia with or without tracheo-oesophagal fistula	Q390 - Q3914	36	7	43	0.2	37	0.2
Duodenal atresia or stenosis	Q410	19	<5	22	0.1	14	0.1
Hirchspung's disease	Q431	27	0	27	0.2	25	0.1
Genital anomalies	Q50 – Q52, Q54 – Q56	**231**	**14**	**245**	**1.4**	**240**	**1.3**
Urinary anomalies	Q60 - Q64, Q794	**322**	**75**	**397**	**2.2**	**379**	**2.1**
Limb anomalies		**361**	**73**	**434**	**2.4**	**404**	**2.3**
Reduction defects	Q71 – Q73	48	24	72	0.4	68	0.4
Club foot – talipes equinovarus	Q660	134	36	170	1.0	160	0.9
Musculo-skeletal, skeletal dysplasias	Q750 – Q751, Q754 –Q759, Q761 – Q764, Q766 – Q769, Q77 – Q78, Q796 – Q799	**76**	**44**	**120**	**0.7**	**117**	**0.7**
Abdominal wall defects		**122**	**75**	**197**	**1.1**	**152**	**0.9**
Diaphragmatic Hernia	Q790	35	15	50	0.3	41	0.2
Gastroschisis	Q793	53	<5	56	0.3	56	0.3
Omphalocele	Q792	34	57	91	0.5	55	0.3
Other anomalies	Q27 – Q28, Q80 – Q85, Q89	**77**	**25**	**102**	**0.6**	**98**	**0.6**
Genetic syndromes & microdeletions	Q87, Q936, D821	**72**	**27**	**99**	**0.6**	**96**	**0.5**
Chromosomal anomalies	Q90 – Q93, Q96 – Q99	**340**	**530**	**870**	**4.9**	**0**	**0.0**
Down's Syndrome (Trisomy 21)	Q90	208	275	483	2.7	0	0.0
Patau syndrome (Trisomy 13)	Q914 – Q917	10	47	57	0.3	0	0.0
Edward syndrome (Trisomy 18)	Q910 – Q913	27	113	140	0.8	0	0.0
Turner's syndrome	Q96	20	34	54	0.3	0	0.0

Part 4 - Information about specific anomalies

1. Open Neural Tube Defects (NTD), year of birth 2005 -2010

Anencephaly: **Definition:** Total or partial absence of the cranial vault, covering skin and brain tissue.

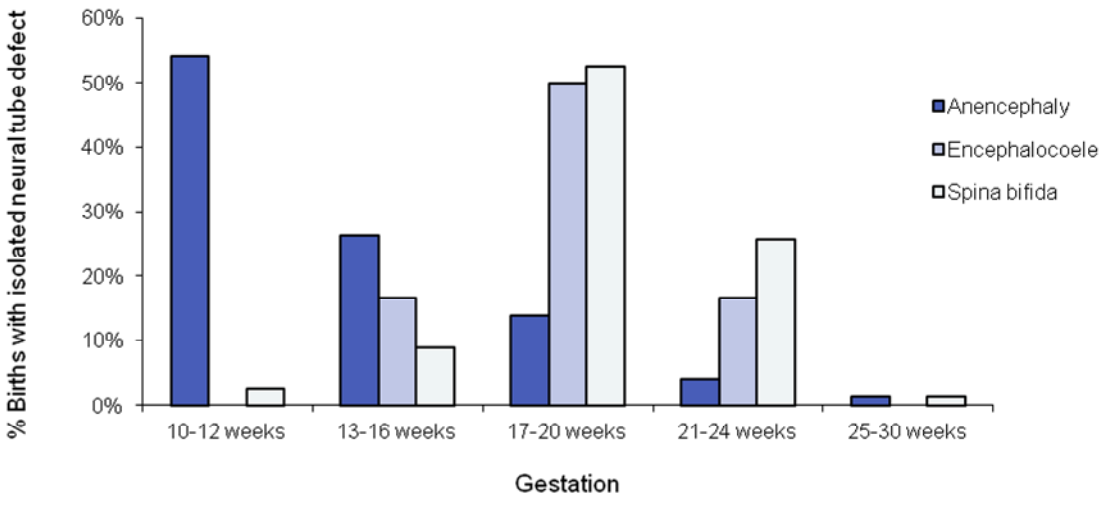
Encephalocoele: **Definition:** Herniation of the brain and/or meninges through a defect in the skull.

Spina bifida: **Definition:** Non-closure of the spine resulting in herniation or exposure of the spinal cord and /or meninges. Hydrocephaly may or may not be present.

Summary Information

Prenatal Investigation:	Ultrasound scan +/- maternal serum alpha feto protein screening
Rate:	
Isolated neural tube defects	0.9 per 1000 births n = 165
Isolated and non-isolated neural tube defects	1.1 per 1000 births n=190
Prenatal detection rate for isolated cases:	157/165 (95%)
ICD 10 codes:	Q00.0 (anencephaly); Q01 – Q01.9 (encephalocoele); Q05 – Q05.9 (spina bifida)

Figure 5 Gestation at prenatal diagnosis of isolated neural tube defects - Percentage of each type (anencephaly, encephalocele, spina bifida) diagnosed at different gestational periods



2. Cardiac Anomalies, year of birth 2005 -2010

Definition: Group of anomalies with abnormal structure of the heart.

Summary information

All Cardiac anomalies

Prenatal Investigation:	Ultrasound scan
Rate:	
Isolated and non-isolated structural cardiac anomalies	4.6[#] per 1000 n = 825
Isolated structural cardiac anomalies	3.2 per 1000 n = 562
Prenatal detection rate of isolated cardiac cases	177/562 (31%)
ICD 10 Codes	Q20 - Q26.9

[#]Expected rate 7-8 per 1,000 (Knowles R et al. Newborn screening for congenital heart defects: a systematic review and cost-effectiveness analysis. *Health Technol Assess* 2005;9(44),1-152)

It has always been recognised that there is under ascertainment of cardiac abnormalities, particularly those diagnosed after the mother has left the maternity unit. In the last two years there has been some improvement due to new outpatient sources. Figure 6 illustrates the prenatal diagnosis rate for some selected isolated cardiac anomalies and Figure 7 the prenatal diagnosis rate for all isolated cardiac anomalies in the 5 year period. The lower rates in 2009 and 2010 probably reflect the improvement in postnatal ascertainment. Figure 8 shows the different type/aetiology for all cardiac cases.

Figure 6 Selected isolated anomalies, number of cases and percentage prenatally diagnosed

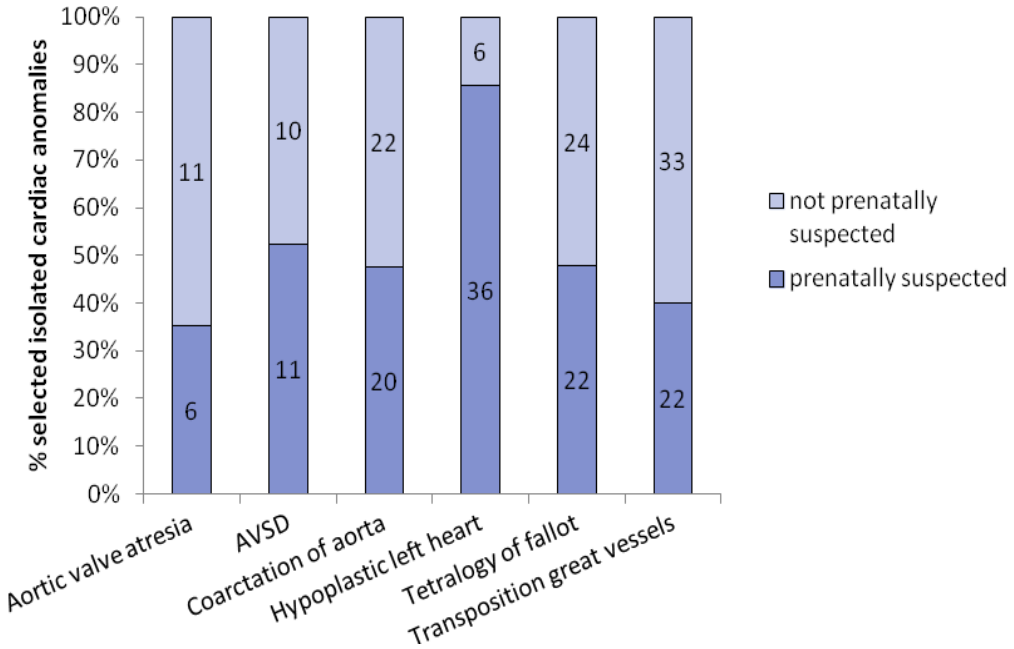


Figure 7 Isolated cardiac anomalies, percentage and number prenatally diagnosed, by year

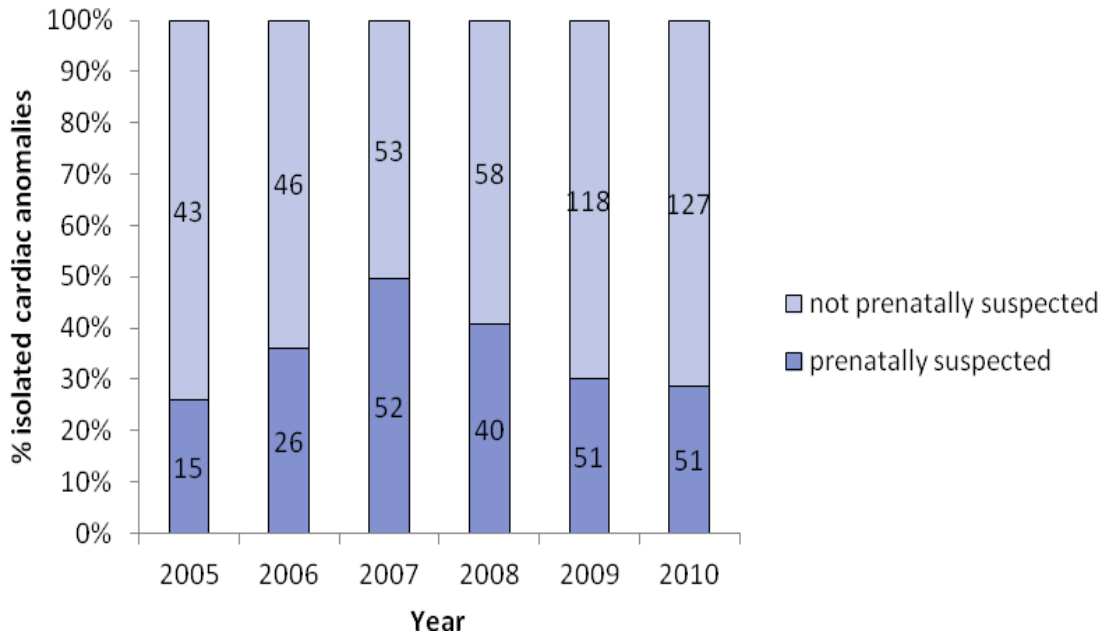
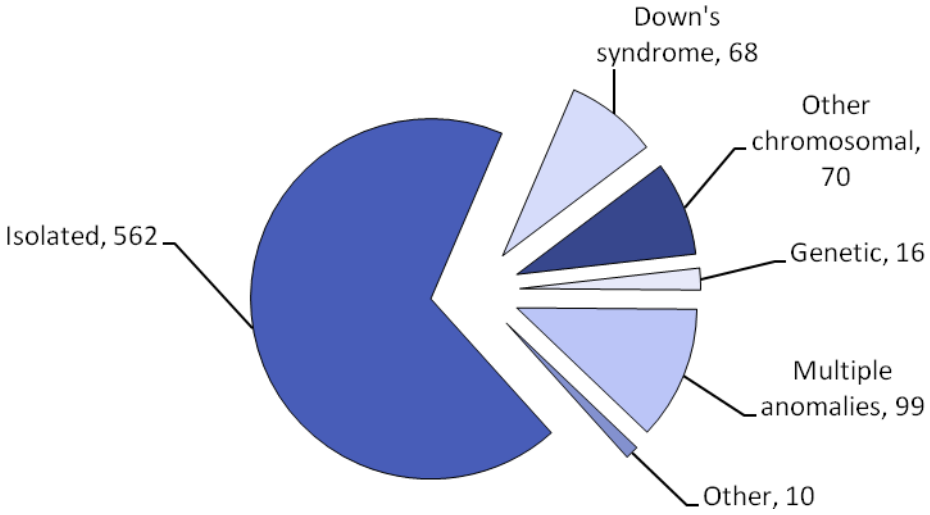


Figure 8 Percentage and number of births with a cardiac anomaly categorised by type, n=825



3. Cleft Lip with or without Cleft Palate (Cleft lip +/- Palate), year of birth 2005 -2010

Cleft lip: **Definition** - Clefting of the upper lip without clefting of the alveolar ridge and palate.

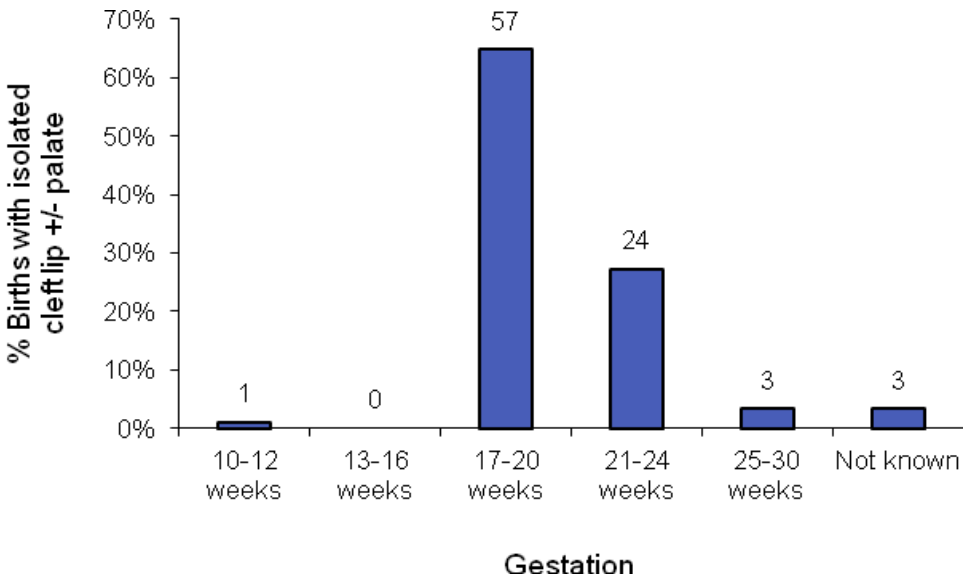
Cleft lip and palate: **Definition** - Clefting of the upper lip with clefting of the alveolar ridge and palate.

Summary Information

Prenatal Investigation:	Ultrasound scan
Rate: **Isolated cleft lip +/- palate**	0.7 / 1,000 n = 126
Prenatal detection rate:	88 / 126 (70%)
ICD 10 Codes	Q36 – 37.9

We report the prenatal detection of cleft lip with or without cleft palate. It is not usually possible to visualise isolated cleft palate by ultrasound prenatally. Very minor clefts (forme fruste) have been excluded from this analysis.
There were 126 cases of isolated cleft lip +/- palate of which 88 (70%) were prenatally diagnosed. There were 40 cases of non-isolated cleft lip +/- cleft palate of which 15 were associated with chromosome anomalies.

Figure 9 Percentage and number of births with prenatally diagnosed isolated Cleft lip +/- palate diagnosed at different gestational age periods, n = 88



4. Diaphragmatic Hernia, Exomphalos and Gastroschisis, year of birth 2005 -2010

a. Diaphragmatic hernia: **Definition -** Herniation of the abdominal organs into the thorax through a defect in the diaphragm.

b. Exomphalos: **Definition -** Herniation of abdominal contents through the umbilical insertion and covered by membrane which may or may not remain intact.

c. Gastroschisis: **Definition -** Visceral herniation through an abdominal wall defect lateral to an intact umbilical cord.

Summary information

	Diaphragmatic Hernia	Exomphalos	Gastroschisis
Prenatal Investigation	Ultrasound scan	Ultrasound scan +/- maternal serum AFP screening	Ultrasound scan +/- maternal serum AFP screening
Number of isolated cases	35	29	55
Non-isolated cases	16 (eg chromosomal, cardiac and renal anomalies)	69 (eg Trisomy 18, Beckwith-Wiedemann syndrome)	1 multiple anomalies
Rate:			
Isolated cases	0.2 / 1,000	0.2 / 1,000	0.3 / 1,000
Isolated and non-isolated cases	0.3 /1,000	0.5 / 1,000	0.3 / 1,000
Prenatal detection rate for isolated cases	24*/35 (69%)	26/29 (90%)	55/55 (100%)
ICD 10 Codes	Q79.0	Q79.2	Q79.3

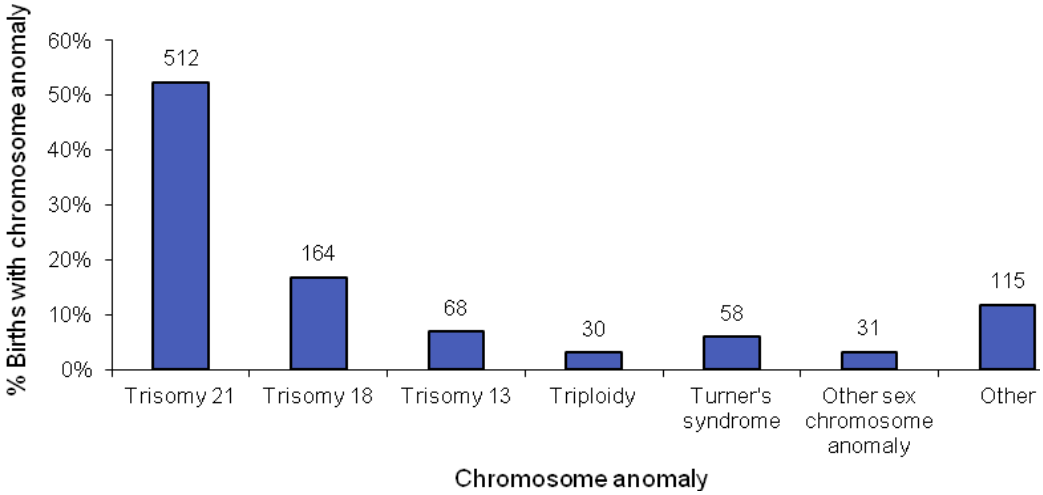
* there were 2 additional cases where there was a suspicion of an anomaly but diaphragmatic hernia was not diagnosed

There was a high prenatal diagnosis rate for cases with isolated gastroschisis (100%) and for isolated exomphalos (90%). Sixty nine percent of isolated diaphragmatic hernia cases had a suspicion on scan prenatally. In one of these cases a cystadenomatous malformation of lung was suspected.

It is well recognised that gastroschisis is more common in babies born to younger mothers and that it is more likely to be an isolated lesion compared to both diaphragmatic hernia and exomphalos. All but 1 of the gastroschisis cases, 69% of diaphragmatic herniae and 30% of exomphalos had isolated lesions in the cases reported to CAROBB and born 2005 – 2010 inclusive. The mean age (range) of mothers babies with gastroschisis was 23 years (17-36 years) compared to 32 years (19-43 years) for isolated exomphalos and 31 years (19-38 years) for isolated diaphragmatic hernia.

5. Chromosome Anomalies, year of birth 2005 -2010

Figure 10 All Chromosome anomalies, percentage of cases and number by chromosome type, n = 978



6. Down's Syndrome (Trisomy 21)

Definition: Additional chromosome 21.

Summary information

Prenatal Investigation:	First and second trimester screening tests. Karyotyping performed because higher risk for Down's syndrome for one of the following reasons: older mother, positive family history, translocation carrier, higher risk screening test or suspicion on ultrasound scan.
Rate:	2.9 / 1,000
From 12 weeks gestation	n = 512
Prenatal detection rate:	305/512 (60%)
ICD 10 Codes	Q90 – Q90.9

Over the last fifteen years there has been a move from offering pregnant women at higher risk for having a baby with Down's syndrome a prenatal diagnostic test, to a national programme for prenatal screening tests to be offered to all pregnant women.

In the CAROBB area there were a variety of screening tests for Down's syndrome in place in 2005 but by 2009 all NHS hospitals were offering first trimester combined screening on the NHS, as recommended by the National Screening Committee Fetal Anomaly Screening Programme www.fetalanomaly.screening.nhs.uk.

There were 512 births with Down's syndrome between 2005 and 2010 inclusive. Two hundred and ninety four (57%) of the 512 cases were karyotyped prenatally before 24 weeks gestation. In 369/512 (72%) of cases there was some prenatal suspicion of abnormality either due to a higher risk screening test result or scan appearance but

karyotyping was not performed in all cases. Figures 11a shows the percentage of Down's syndrome cases prenatally diagnosed, those with some prenatal suspicion and those with no suspicion prenatally, by year. Figure 11b shows the percentage of cases prenatally diagnosed at different gestational ages, by year. These show a tendency towards a higher prenatal diagnosis rate and earlier gestation at diagnosis.

Fig 11a Percentage of Downs Syndrome cases prenatally diagnosed, percentage with some prenatal suspicion, and percentage with no prenatal suspicion, by year (n=512)

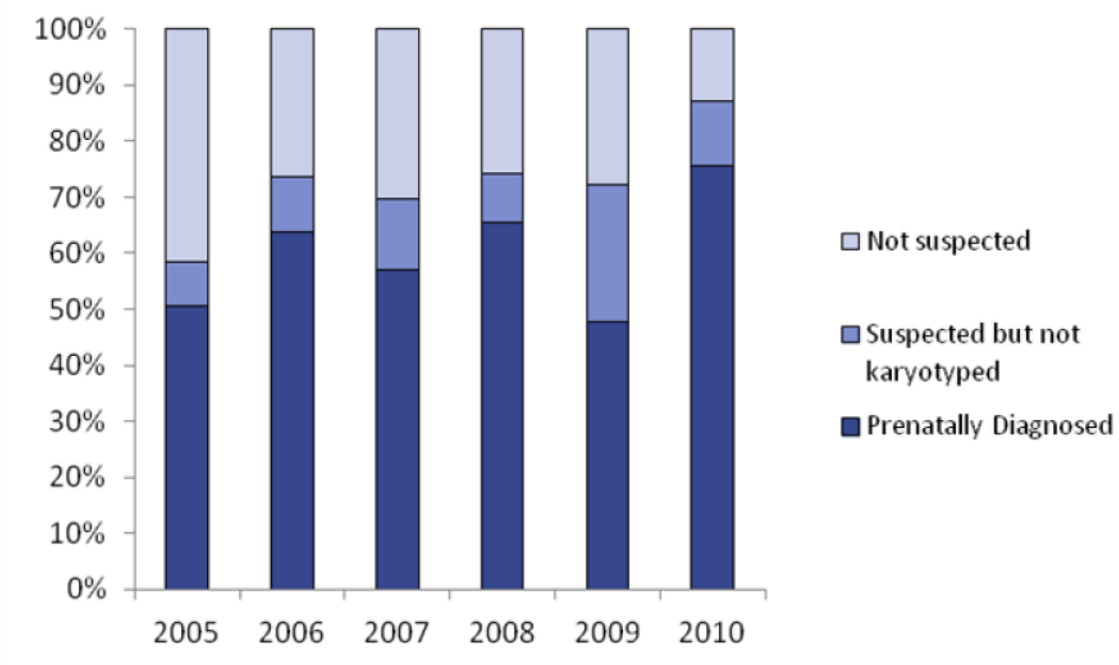
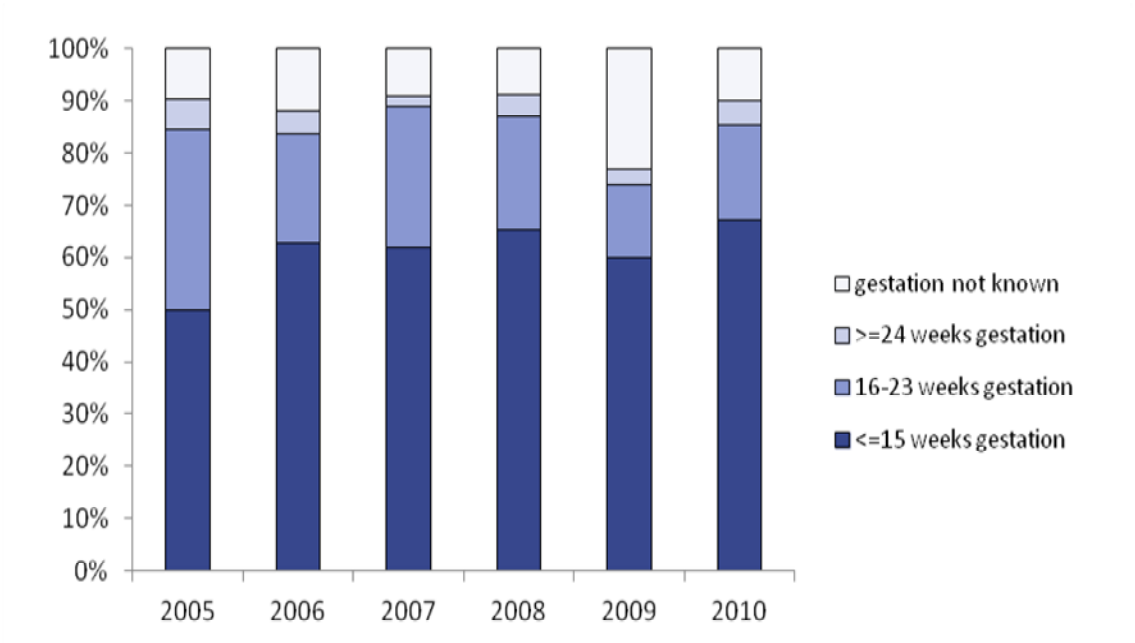


Fig 11b Percentage of prenatally suspected Down's syndrome cases diagnosed at different gestational ages, by year (n=369)



Appendices

Congenital Anomalies from an unselected population within Oxfordshire, 1991-2010 using data from OXCAR and CAROBB

Summary table

Table 1A: Prenatal detection of selected congenital anomalies from an unselected population within Oxfordshire, 1991 – 2010

Defect	Prenatal investigation	Number of pregnancies notified with prenatal suspicion of anomaly (not including false positive diagnoses)	Number of cases notified with anomaly confirmed at birth	Prevalence per 1,000 total births	Prenatal detection rate
Isolated open neural tube defects (anencephaly & spina bifida)	Ultrasound Scanning +/- MS AFP	146	156	1.2	94%
Isolated cardiac anomaly	Ultrasound scanning	145	471	3.6	31%
Isolated cleft lip +/- palate	Ultrasound scanning	59	95	0.7	62%
Down's syndrome	Karyotyping Prenatal detection because MA>35 or 1st or 2nd trimester screening test or ultrasound scanning	245 (195 karyotyped)	356	2.8	69% (55%)
Isolated diaphragmatic hernia	Ultrasound scanning	22	36	0.3	61%
Isolated exomphalos (excludes exomphalos minor)	Ultrasound scanning +/- MS AFP	27	31	0.2	87%
Isolated gastroschisis	Ultrasound scanning +/- MS AFP	32	32	0.2	100%

[1] There is under reporting of cardiac anomalies diagnosed after discharge from the maternity unit particularly for years 1991-2007

Background

The Oxford Congenital Anomaly Register (OXCAR) was established 21 years ago, in 1991, after consultation with local experts (obstetricians, midwives, paediatricians, neonatologists, paediatric cardiologists, paediatric pathologists, geneticists, biochemists and public health physicians) who gave full support to the register. One of the main aims of the register at that time was to monitor the newly developing techniques used in prenatal diagnosis and particularly the accuracy of antenatal ultrasound scanning. The first six years of data were summarised in a paper published in the Lancet in 1998 (see Appendix 4 reference 42).

Other aims were to improve ascertainment to the then National Congenital Anomaly System for surveillance (now carried out by BINOCAR), to provide data for health care policies and planning and for research on aetiology and natural history of congenital anomalies to enable better advice to be given to parents and prospective parents. In 2003 funding from the Department of Health enabled the expansion of OXCAR to Berkshire and Buckinghamshire (i.e. to cover Thames Valley) and the name was changed to CAROBB. Because there is now 20 years of data for the Oxford area, we are, in this

Appendix 1

Appendix to the main CAROBB report, summarising these data. More detailed information is available about individual anomalies, prenatal detection rates and outcome of pregnancy. Please contact us by email at carobb@npeu.ox.ac.uk if you would like further information.

The population studied

Anomalies suspected and or confirmed in fetuses / babies booked for delivery at the Oxford Women's Centre, John Radcliffe Hospital, community hospital or home delivery within the catchment area of the Women's Centre and with an OX postcode during 1991 - 2010 inclusive. Denominator data for this population were provided by the Oxford Radcliffe Hospitals NHS Trust Performance & Information Department. There were 129163 births in this category in the 20 year study period. Please note this population does not equate with the data from the whole of Oxfordshire used in the CAROBB report. The population used here gives the best approximation available to the unselected local Oxford population.

Table 2A: Total births and notifications from an unselected population within Oxfordshire, (John Radcliffe Women's Centre booking, with OX postcodes), 1991-2010 inclusive; number prenatally suspected with and without congenital anomaly at birth, number resulting in termination of pregnancy for fetal anomaly (TOPFA), in six three-year periods

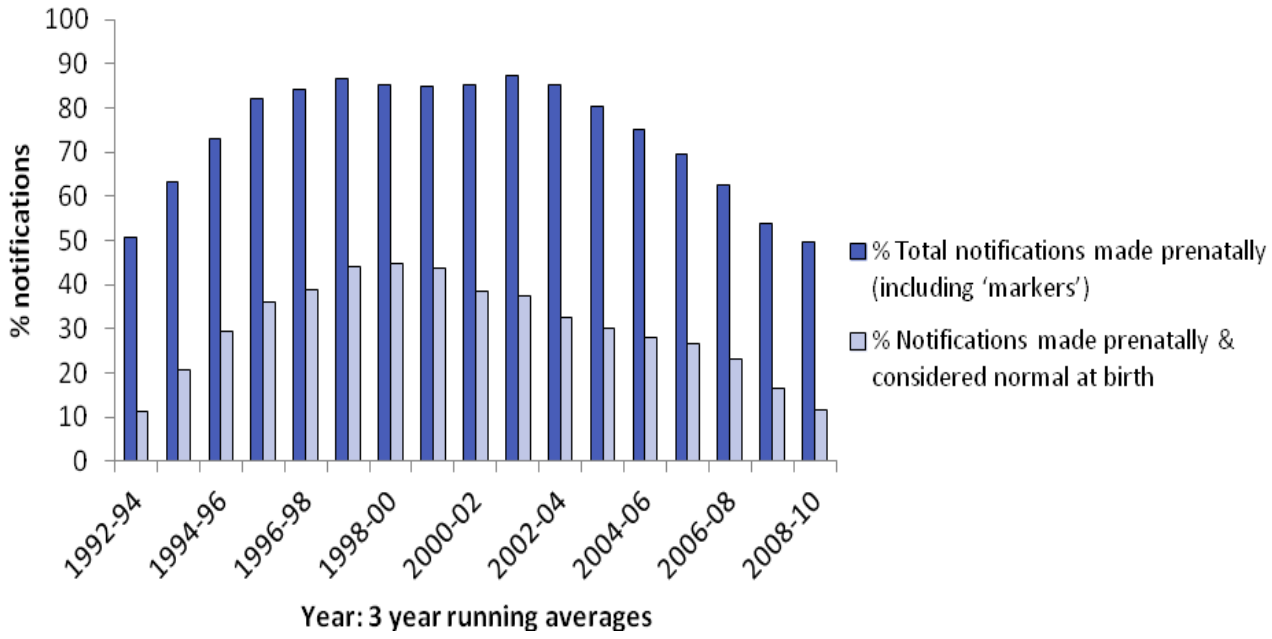
Year	1991-1995	1996-2000	2001-2005	2006-2010	**1991-2010**
Total births	28966	29120	33348	37729	**129163**
Total notifications	729	1219	1020	1461	**4429**
Total notifications made prenatally (including 'markers') **(% of total notified)**	421 (58%)	1038 (85%)	843 (83%)	826 (57%)	**3128 (71%)**
Notifications made prenatally with anomaly at birth **(% of total)**	289 (40%)	521 (43%)	499 (49%)	564 (39%)	**1873 (42%)**
Notifications made prenatally & considered normal at birth **(% of total notified prenatally)**	132 (31%)	514 (50%)	340 (40%)	256 (31%)	**1242 (40%)**
Notifications made prenatally and resulting in TOPFA **(% of prenatally diagnosed cases with anomaly confirmed)**	128 (44%)	216 (44%)	211 (42%)	242 (43%)	**797 (43%)**
Total with anomaly at delivery. **(% of total births)**	597 (2.1%)	702 (2.4%)	676 (2.0%)	1199 (3.2%)	**3174** (2.5%)
Proportion of total births with prenatal suspicion & baby normal birth*	1 in 219	1 in 57	1 in 98	1 in 147	**1 in 104**

< 1% lost to follow up

Table 2A gives the number of notifications to the OXCAR population in four five-year periods from 1991 – 2010. During these time periods the percentage of cases notified prenatally changed from 58% in the first five years (1991 – 1995), to 85 / 83% in the middle time period (1996-2005) and dropped to 57% during 2006-2010. The apparent fall in the percentage of anomalies detected prenatally (from 49% in 2001-2005 to 39% in 2006-2010) is probably due to improvement in the ascertainment of postnatally diagnosed anomalies due to new sources of ascertainment – particularly for cardiac anomalies. In the same time periods the number of cases where there was a prenatal suspicion but the baby was apparently normal at birth rose from 18% of prenatal notifications in 1991 – 1995 to 42% in 1996-2000 but dropping back to 18% for the years 2006-20010.

This trend is illustrated in Figure 2A which, using 3 year running averages shows the percentage of notification made prenatally and those considered to be normal at birth. This demonstrates the evolution of reporting ultrasound soft markers (normal variants) such as echogenic bowel and nuchal thickening. These started to be reported regularly in the early 1990s. By the mid-1990s it was realised that most babies with these usually normal variants were normal. Local protocols were drawn up to guide professionals on the management of such markers, when to report specific markers and what further tests might be indicated and in 2009 the Fetal Anomaly Screening Programme (FASP) produced national guidelines concerning how to manage the reporting of ultrasound normal variants. www.fetalanomaly.screening.nhs.uk/standardsandpolicies.

Figure 2A Percentage of notification made prenatally and those considered normal at birth using 3 year running averages



Appendix 2

CAROBB Notification form

The standard notification form is shown overleaf but we are happy to accept information in other ways eg copies of discharge letters or clinic lists.

Please contact us if you would like to discuss how best to notify to the register.

We will provide copies of forms on request or forms can be printed from our website: www.npeu.ox.ac.uk/carobb

CAROBB NOTIFICATION FORM

Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire

Office use only - Case no

Dup | Com | From

Please register any actual OR prenatally suspected anomaly - structural, chromosomal or biochemical in fetus/baby. (See reverse of form for more information about the register and exclusion list)

MOTHER DETAILS

(Sticky label, if available)

Surname.....

Forename.....**Hosp No**.....

NHS Number.....

Postcode *(essential field)*

Mother's DoB *(essential field)*

Booking hosp.....

To deliver at.....
(if different from booking hospital)

EDD *(essential field)*

Mother BMI.....

Assisted conception / IVF?..... *If yes, (please state method, if known)*

No of previous pregnancies/births

.....Livebirth .....Miscarriage/TOP *(<24 weeks)* .....Stillbirth/TOP *(>24 weeks)*

Ethnic origin of mother *(please circle)*

White	Pakistani	Black Caribbean	Chinese
Mixed	Bangladeshi	Black African	Other
Indian	Other Asian	Other Black	Not known

BABY DETAILS

(Sticky label, if available)

Surname.....

Forename.....**Hosp No**.....

NHS Number.....

Sex Male / Female / Ambiguous / Not known
(please circle)

Date of delivery / TOP
(and date of feticide if performed)

Place of delivery.....

Gest at delivery.....**weeks**

Weight g ☐ Not weighed

Multiple pregnancy?.....**Zygosity:** MCMA / MCDA / DCDA

Outcome *(when possible, please report date of delivery, gest, sex, weight and details of any anomalies, whatever the outcome)*

- ☐ Liveborn, no anomaly identified, no follow up requested
- ☐ Liveborn, anomaly present or req' further tests *(please give details)*
- ☐ Miscarriage/IUD *(<24 weeks)*
- ☐ Stillbirth/IUD *(>24 weeks)*
- ☐ Termination
- ☐ Neonatal death

Date of neonatal death

Post mortem? Yes / No / Not known

PRENATAL INVESTIGATIONS

Screening and Diagnostic tests

Gest	Test *(please circle)*	Result
.....	Nuchal / Combined	NT measurement .....mm
.....	Triple	Down's risk 1 in .....
.....	Other	Tri 13 / 18 risk 1 in .....
		Not offered / Declined
.....	CVS / Amnio	Normal / Abnormal *(state karyotype if known)*
.....	FISH / PCR	.....
		Not offered / Declined
.....	Other *(please state)*	

Gest	Ultrasound scan findings *(& any other relevant details)*
.....	.....
.....	.....
.....	.....
.....	.....
.....	.....
.....	.....

POSTNATAL DETAILS OF ANOMALY

Prenatally suspected? ☐ Yes ☐ No

.....

.....

.....

.....

.....

.....

Surgery? Performed / Expected in 1st year / Expected after 1st year

Additional details *(eg previous congenital anomalies, illness in mother, exposure to potentially harmful substances)*

.....

.....

.....

.....

Referred to:.....

Consanguinity? 1st cousins / other relation.....
(if applicable, please circle / state)

Notified by:.....Date:.....Hospital:.....Dept:.....Tel:.....

Confidential: Please send in a sealed envelope to: CAROBB, NPEU, RDB, Old Road Campus, Oxford OX3 7LF or use confidential fax: 01865 289720. Any queries contact Cath Rounding: Tel: 01865 289721, E-mail: CAROBB@npeu.ox.ac.uk.

Appendix 2

Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire (CAROBB)

Please complete the form overleaf as fully as possible, registering any anomalies found at whatever stage you become aware of them in the pregnancy/postnatal period.

Uses of the register:

- Audit for prenatal diagnosis
- Evaluation and monitoring of new invasive and non invasive prenatal tests
- Evaluation of new screening programmes
- Provision of data for health care policies and planning
- Provision of data for the investigation of cluster of abnormalities
- Investigation of putative teratogens
- Research on aetiology and natural history of particular malformations
- Improving ascertainment to the National Congenital Anomaly System

Congenital anomalies exclusion list
It is not necessary to report any of the following conditions to us POSTNATALLY, unless there was a prenatal suspicion of an anomaly.

- Spina bifida occulta uncomplicated
- Phymosis
- Stenosis or stricture of lacrimal duct
- Minor skin anomalies less than $4cm^2$: skin tag, naevus, angioma, haemangioma, glomus tumor, lymphangioma, birth mark
- Minor anomaly of auricle
- Clicking hip
- Minor anomaly of face or nose
- Minor anomaly of nipple, accessory or ectopic nipple
- Postural clubfoot
- Minor anomalies of the foot: hallux valgus/varus, "orteil en marteau", metatarus valgus/adductus
- Postural talipes calcaneovalgus or pes calcaneovalgus
- Congenital umbilical hernia, inguinal or para umbilical
- Functional or unspecified cardiac murmur
- Absence or hypoplasia of umbilical artery
- Congenital hydrocele or hydrocele of testis

If in doubt, report to us, we will feed back any inappropriate reporting

Confidentiality and data protection

All information held on the register is strictly confidential. Data are stored in a secure environment at the National Perinatal Epidemiology Unit, University of Oxford (data protection registration number: Z575783X). Any research undertaken is subject to ethical approval. The register holds Patient Information Advisory Group approval.

Confidential: Please fax or send in a sealed envelope to:

Cath Rounding
CAROBB Co-ordinator
National Perinatal Epidemiology Unit
University of Oxford,
Old Road Campus
Headington
Oxford OX3 7LF

Confidential fax: 01865 289720

Please do not hesitate to contact us with any queries, or requests for more forms.

Tel: 01865 289721
E-mail: carobb@npeu.ox.ac.uk
catherine.rounding@nhs.net
Website: http://www.npeu.ox.ac.uk/carobb/

PLEASE DO NOT SEND ANY NOTIFICATIONS BY E-MAIL, UNLESS USING NHS.NET

Research Projects using data from CAROBB

Ongoing projects

1.	**Project title:**	Gastroschisis
	Investigators:	Elizabeth Draper
	Collaboration:	BINOCAR
	Status of study:	Ongoing
2.	**Project title:**	Sentinel phenotypes
	Investigators:	Ms Suzhuang Hong, Helen Dolk, Marlene Sinclair, Diana Wellesley, ingeborg Barisic, Maria Loane, Ian Bradbury
	Collaboration:	EUROCAT
	Status of study:	Ongoing
3.	**Project title:**	Fraser Syndrome
	Investigators:	Helen Dolk, Ingeborg Barisic
	Collaboration:	EUROCAT
	Status of study:	Ongoing
4.	**Project title:**	Esophageal Atresia: Population based study of Epidemiology and outcome in European Regions.
	Investigators:	Rikke Neess Pedersen, Ester Garne, Steffen Husby
	Collaboration:	EUROCAT
	Status of study:	Ongoing
5.	**Project title:**	The Risk of Congenital Anomalies in Multiple Births: A European Registry Based Study
	Investigators:	Breidge Boyle
	Collaboration:	EUROCAT
	Status of study:	Ongoing
6.	**Project title:**	Prevalence of neural tube defects (NTD) in younger mothers in Europe 2000-2008: analysis of the EUROCAT database
	Investigators:	M Loane, H Dolk, J Morris, H de Walle, L Abramsky & EUROCAT Working Group
	Collaboration:	EUROCAT
	Status of study:	Ongoing
7.	**Project title:**	Association between specific congenital heart anomalies and Smith Lemli Opitz like birth defects
	Investigators:	ME Smilde-Baardman, MK Bakker, WS Kerstjens-Frederikse, RMW Berger & EUROCAT Working Group
	Collaboration:	EUROCAT
	Status of study:	Ongoing
8.	**Project title:**	Trends and patterns of sirenomelia and cyclopia in Europe, a descriptive study based on the European surveillance system of congenital anomalies (EUROCAT)
	Investigators:	Harry Pachajoa, Carolina Isaza, Fabian Mendez
	Collaboration:	EUROCAT
	Status of study:	Ongoing

Appendix 3

9.	**Project title:**	Termination of pregnancy for non lethal fetal anomaly: professional perspectives.
	Investigators:	Lisa Crowe, Ruth Graham, Judith Rankin, Steve Robson
	Collaboration:	BINOCAR
	Status of study:	Ongoing
10.	**Project title:**	Antenatal diagnosis of lissencephaly
	Investigators:	Paul Griffiths, Mike Reeves
	Collaboration:	BINOCAR
	Status of study:	Ongoing
11.	**Project title:**	Total & livebirth prevalence of Down syndrome and other trisomies in Europe 1990-2007: impact of increasing maternal age, prenatal screening and termination of pregnancy
	Investigators:	Maria Loane, Helen Dolk, Joan K Morris, Marie-Claude Addor, Larraitz Arriola,Berenice Doray, Patricia Boyd, Elizabeth Draper or Judith BuddEster Garne, Miriam Gatt, Martin Haeusler, Babak Khoshnood, et al
	Collaboration:	EUROCAT
	Status of study:	Ongoing
12.	**Project title:**	Children with language, reading and communication problems
	Investigators:	Dorothy Bishop, Debbie Shears, Patricia Boyd
	Collaboration:	Local
	Status of study:	Ongoing
13.	**Project title:**	Investigating the epidemiology of partial urorectal septum malformation sequence: a population-based study using data from the British Isles Network fo Congenital Anomaly Registers (BINOCAR)
	Investigators:	Judith Rankin, Peter Tennant, Svetlana Glinianaia, Diana Wellesley
	Collaboration:	BINOCAR
	Status of study:	Ongoing
14.	**Project title:**	The evolution of prenatal screening and diagnosis and its impact on an unselected population over an 18 year period
	Investigators:	Patricia Boyd
	Collaboration:	Local
	Status of study:	Ongoing
15.	**Project title:**	Epidemiology of Hirschsprung's disease in Europe: a register-based study
	Investigators:	Judith Rankin, Kate Best
	Collaboration:	EUROCAT
	Status of study:	Ongoing
16.	**Project title:**	Epidemiology of Rare Syndromes in Europe
	Investigators:	Helen Dolk, Ingeborg Barisic
	Collaboration:	EUROCAT
	Status of study:	Ongoing

17.	**Project title:**	Investigating the association between congenital anomalies and childhood cancer: a population-based data- linkage study
	Investigators:	Judith Rankin, Peter Tennant
	Collaboration:	BINOCAR
	Status of study:	Ongoing
18.	**Project title:**	Epidemiology of orofacial clefts and associated malformations in a geographically defined region.
	Investigators:	Jenaleen Law
	Collaboration:	Local
	Status of study:	Ongoing
19.	**Project title:**	The impact of prenatal screening and susbsequent terminations on the prevalence of CHD anomalies in live born babies with Down syndrome
	Investigators:	Prof Joan Morris, Ester Garne, Diana Wellesley, Anna Springett
	Collaboration:	EUROCAT
	Status of study:	Ongoing
20.	**Project title:**	Epidemiology of congenital diaphragmatic hernia in Europe: a register-based study
	Investigators:	Judith Rankin, Mark McGivern, Kate Best
	Collaboration:	EUROCAT
	Status of study:	Ongoing

Appendix 3

Completed projects and one-off data requests

21.	**Project title:**	Congenital hydrocephalus: a population based study on prevalence and outcome
	Investigators:	Ester Garne
	Collaboration:	EUROCAT
	Status of study:	Complete
22.	**Project title:**	Chlorination of water supplies and birth defects
	Investigators:	Paul Elliott
	Collaboration:	SASHU
	Status of study:	Complete
23.	**Project title:**	Local investigation of potential cluster
	Investigators:	G Dean
	Collaboration:	Local
	Status of study:	Complete
24.	**Project title:**	Investigation of neural tube defects near landfill site
	Investigators:	Nick Hicks
	Collaboration:	Local
	Status of study:	Complete
25.	**Project title:**	NCAS alert re cardiac & urogenital anomalies
	Investigators:	Monica Dent
	Collaboration:	Other
	Status of study:	Complete
26.	**Project title:**	Concern from member of public re rise in no of anomalies since 1995
	Investigators:	Don Sinclair
	Collaboration:	Local
	Status of study:	Complete
27.	**Project title:**	Clinical genetics audit of late TOP
	Investigators:	Dorothy Halliday, Patricia Boyd
	Collaboration:	Local
	Status of study:	Complete
28.	**Project title:**	Geographical variation in overall rates of congenital abnomalities and the rates for specific abnormalities
	Investigators:	Helen Dolk
	Collaboration:	EUROCAT
	Status of study:	Complete
29.	**Project title:**	Myotonic dystrophy audit
	Investigators:	Paul Chamberlain
	Collaboration:	Local
	Status of study:	Complete
30.	**Project title:**	How have babies born with spina bifida in the 1990's fared?
	Investigators:	Jenny Kurinczuk, Jenny Calvert, Patricia Boyd, Paul Chamberlain, Mary Anthony

	Collaboration:	Local
	Status of study:	Complete
31.	**Project title:**	Follow-up Of Children with Congenital Anomalies Long-term. (FOCAL) Pilot study of diaphragmatic hernia
	Investigators:	FOCAL
	Collaboration:	BINOCAR & BDF Newlife
	Status of study:	Complete
32.	**Project title:**	Absent stomach bubble/TOF/OA
	Investigators:	Paul Chamberlain, Kokila Lakhoo, Patricia Boyd
	Collaboration:	Local
	Status of study:	Complete
33.	**Project title:**	Understanding congenital anomaly hotspots within Oxon (postcode mapping)
	Investigators:	Angela Baker
	Collaboration:	Local
	Status of study:	Complete
34.	**Project title:**	Audit of screening of fetuses with echogenic bowel
	Investigators:	Gail Whitehead
	Collaboration:	Local
	Status of study:	Complete
35.	**Project title:**	Audit of screening offered to parents of those babies born with Down's syndrome
	Investigators:	Gail Whitehead
	Collaboration:	Local
	Status of study:	Complete
36.	**Project title:**	Survey of congenital lung anomalies
	Investigators:	Mary Anthony
	Collaboration:	
	Status of study:	Complete
37.	**Project title:**	Assessment of ultrasound markers and their value
	Investigators:	National Screening Committee
	Collaboration:	Local
	Status of study:	Complete
38.	**Project title:**	Arthrogryposis multiplex congenita (AMC) – causes and risk factors
	Investigators:	Jana Midelfart Hoff
	Collaboration:	EUROCAT
	Status of study:	Complete
39.	**Project title:**	Audit of gastroschisis 1995-2005
	Investigators:	Gail Whitehead
	Collaboration:	Local
	Status of study:	Complete
40.	**Project title:**	Audit of prenatal lung lesions versus pathological diagnosis

Appendix 3

	Investigators:	P Teong, K Lakhoo, L Impey
	Collaboration:	Local
	Status of study:	Complete
41.	**Project title:**	Incidence of brain anomalies
	Investigators:	Marion Knight
	Collaboration:	Other
	Status of study:	Complete
42.	**Project title:**	Isolated cleft lip and palate audit
	Investigators:	Dorothy Halliday, Patricia Boyd
	Collaboration:	Local
	Status of study:	Complete
43.	**Project title:**	Prenatal screening in Europe
	Investigators:	Patricia Boyd, Ester Garne
	Collaboration:	EUROCAT
	Status of study:	Complete
44.	**Project title:**	Oro-facial Clefts. World-wide Recent Total Prevalence Data.
	Investigators:	Pierpaolo Mastroiacovo
	Collaboration:	Other
	Status of study:	Complete
45.	**Project title:**	Antenatal diagnosis of duodenal atresia and postnatal outcome
	Investigators:	Ms PG Roy, K Lakhoo, P Boyd
	Collaboration:	Local
	Status of study:	Complete
46.	**Project title:**	Maternal age-specific risk of non-chromosomal anomalies
	Investigators:	M Loane, H Dolk, JK Morris, EUROCAT Working Group
	Collaboration:	EUROCAT
	Status of study:	Complete
47.	**Project title:**	Terminations of pregnancy ≥ 24 weeks of gestation after prenatal diagnosis of fetal abnormality in Europe
	Investigators:	Ester Garne, Helen Dolk, Patricia Boyd, Maria Loane, Catherine de Vigan, Babak Khoshnood
	Collaboration:	EUROCAT
	Status of study:	Complete
48.	**Project title:**	The epidemiology of hospital admission rates for cleft lip and palate in the United Kingdom
	Investigators:	Aadil A Khan, Tim Goodacre
	Collaboration:	Local
	Status of study:	Complete
49.	**Project title:**	Sex Chromosome Trisomies in Europe: Prevalence, prenatal detection and outcome of pregnancy
	Investigators:	PA Boyd, M Loane, E Garne, B Khoshnood, H Dolk, and a EUROCAT working group
	Collaboration:	EUROCAT

	Status of study:	complete

50.	**Project title:**	Cornelia de Lange Syndrome
	Investigators:	Helen Dolk, Ingeborg Barisic
	Collaboration:	EUROCAT
	Status of study:	Complete

51.	**Project title:**	Cognitive and behavioural outcomes of children with an extra sex chromosome
	Investigators:	Pat Jacob, Dorothy Bishop, Gaia Scerif
	Collaboration:	Dept of Experimental Psychology, Oxford University; Wessex Regional Genetics Laboratory
	Status of study:	Complete

52.	**Project title:**	To define the outcome of prenatally diagnosed gastroschisis with intra abdominal bowel dilatation vs those with no dilatation in the Thames Valley Region
	Investigators:	Kokila Lakhoo
	Collaboration:	Local
	Status of study:	Complete

53.	**Project title:**	A descriptive epidemiological study of small intestinal atresia in Europe
	Investigators:	Judith Rankin
	Collaboration:	EUROCAT
	Status of study:	Complete

54.	**Project title:**	Evaluation of prenatal diagnosis rates for major structural congenital anomalies across areas covered by the British Isles Network of Congenital Anomaly Registers: 2005 to 2006
	Investigators:	Patricia A Boyd, Ann M Tonks, Judith Rankin, Catherine Rounding, Diana Wellesley, Elizabeth S Draper, and the BINOCAR working group
	Collaboration:	BINOCAR
	Status of study:	Complete

55.	**Project title:**	BINOCAR downs syndrome prenatal screening audit
	Investigators:	
	Collaboration:	BINOCAR
	Status of study:	Complete

56.	**Project title:**	Analysing the rare unbalanced chromosome abnormalities reported to EUROCAT
	Investigators:	Diana Wellesley, Ingeborg Barisic, Patricia Boyd, Helen Dolk, Ruth Greenlees
	Collaboration:	EUROCAT
	Status of study:	Complete

57.	**Project title:**	The outcomes of antenatally diagnosed isolated heart anomalies
	Investigators:	Moira Blyth and Diana Wellesley
	Collaboration:	Inter-register
	Status of study:	Complete

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58.	**Project title:**	Report on the data collected on congenital anomalies in South East Region for surveillance and for monitoring the national antenatal Down's syndrome and fetal anomaly screening programmes.
	Investigators:	Val Armstrong, Patricia A Boyd, Diana Wellesley and Catherine Rounding
	Collaboration:	Inter-register
	Status of study:	Complete
59.	**Project title:**	Audit of known fetal abnormalities communicated to neonatologists for CNST Standard 5
	Investigators:	Mary Anthony
	Collaboration:	Local
	Status of study:	Complete
60.	**Project title:**	Ambient air pollution and risk of congenital anomalies in England, 1991-99
	Investigators:	Dolk H, Armstrong B, Lachowycz K, Vrijheid M, Rankin J, Abramsky L, Boyd PA, Wellesley D
	Collaboration:	EUROCAT
	Status of study:	Complete
61.	**Project title:**	Survey of congenital diaphragmatic hernia
	Investigators:	Mary Anthony, Spr Vikranth Venugopalan
	Collaboration:	Local
	Status of study:	Complete
62.	**Project title:**	Second report of the Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire (CAROBB) CAROBB births 2005-2008 and Oxford births 1991-2008
	Investigators:	Patricia Boyd, Catherine Rounding, Jennifer Kurinczuk
	Collaboration:	Local
	Status of study:	Complete
63.	**Project title:**	EUROCAT Website data on prenatal detection rates of congenital anomalies.
	Investigators:	Ester Garne, Helen Dolk, Maria Loane, Patricia Boyd on behalf of EUROCAT
	Collaboration:	EUROCAT
	Status of study:	Complete
64.	**Project title:**	Data exchange with Down's register
	Investigators:	Cath Rounding, Joan Morris
	Collaboration:	Inter-register
	Status of study:	Complete
65.	**Project title:**	Congenital Heart Defects in Europe: Prevalence and Perinatal Mortality
	Investigators:	Helen Dolk, Maria Loane, Ester Garne.
	Collaboration:	EUROCAT
	Status of study:	Complete

66.	**Project title:**	NTD figures for England and Wales
	Investigators:	Elizabeth Draper
	Collaboration:	BINOCAR
	Status of study:	One off data request
67.	**Project title:**	Sacrococcygeal teratoma audit
	Investigators:	Kokila Lakhoo
	Collaboration:	Local
	Status of study:	One off data request
68.	**Project title:**	AVSD audit
	Investigators:	Paul Chamberlain
	Collaboration:	Local
	Status of study:	One off data request
69.	**Project title:**	Prenatally suspected heart defects in down's syndrome
	Investigators:	Nick Archer
	Collaboration:	Local
	Status of study:	One off data request
70.	**Project title:**	Annual report of anomalies to feedback to antenatal department
	Investigators:	Ann Folkes
	Collaboration:	Local
	Status of study:	One off data request
71.	**Project title:**	Risk management review of cleft lips/palates
	Investigators:	Michelle Errington
	Collaboration:	Local
	Status of study:	One off data request
72.	**Project title:**	Audit cystic hygroma and neonatal outcome
	Investigators:	Kokila Lakhoo
	Collaboration:	Local
	Status of study:	One off data request
73.	**Project title:**	Gastroschisis rates for 2002-2006 for TVSHA to compare with SWCAR
	Investigators:	Aileen McLoughlin
	Collaboration:	Local
	Status of study:	One off data request
74.	**Project title:**	Supply of data for National Screening Committee - cases of anencephaly and gastroschisis
	Investigators:	Anne Roberts
	Collaboration:	Local
	Status of study:	One off data request
75.	**Project title:**	Supply of data for National Screening Committee - Down's cases
	Investigators:	Anne Roberts
	Collaboration:	Local
	Status of study:	One off data request

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76.	**Project title:**	Audit of craniofacial anomalies
	Investigators:	Paul Chamberlain
	Collaboration:	Local
	Status of study:	One off data request
77.	**Project title:**	Gastroschisis case matching exercise for UKOSS
	Investigators:	Marian Knight
	Collaboration:	Other
	Status of study:	One off data request
78.	**Project title:**	Outcome of prenatally diagnosed exomphalos
	Investigators:	Kokila Lakhoo, N Shenker, J Sadiq
	Collaboration:	Local
	Status of study:	One off data request
79.	**Project title:**	Abdominal cyst audit
	Investigators:	Kokila Lakhoo
	Collaboration:	Local
	Status of study:	One off data request
80.	**Project title:**	Echogenic bowel audit - cross referencing of cases
	Investigators:	Jackie Lovstrom
	Collaboration:	Local
	Status of study:	One off data request
81.	**Project title:**	Gastroschisis case matching exercise with UKOSS
	Investigators:	Marian Knight
	Collaboration:	Other
	Status of study:	One off data request
82.	**Project title:**	Prevalence of CCAM and other thoracic anomalies
	Investigators:	Steve Gould
	Collaboration:	Local
	Status of study:	One off data request
83.	**Project title:**	Exomphalos audit
	Investigators:	Elizabeth Draper
	Collaboration:	BINOCAR
	Status of study:	One off data request
84.	**Project title:**	Gastroschisis audit
	Investigators:	Elizabeth Draper
	Collaboration:	BINOCAR
	Status of study:	One off data request
85.	**Project title:**	Bladder exstrophy cases – numbers prenatally detected.
	Investigators:	Diana Wellesley
	Collaboration:	Inter-register
	Status of study:	One off data request
86.	**Project title:**	Gastroschisis numbers 2002-08.
	Investigators:	Kokila Lakhoo

	Collaboration:	Local
	Status of study:	One off data request
87.	**Project title:**	Audit of soft markers in a population already screened for aneuploidy in the first trimester.
	Investigators:	Lawrence Impey
	Collaboration:	Local
	Status of study:	One off data request
88.	**Project title:**	Comparative incidence and prevalence of abdominal wall defects
	Investigators:	Kokila Lakhoo
	Collaboration:	Local
	Status of study:	One off data request
89.	**Project title:**	Schizencephaly
	Investigators:	David Howe
	Collaboration:	BINOCAR
	Status of study:	One off data request
90.	**Project title:**	TOF cases – data exchange with UKOSS
	Investigators:	Marian Knight
	Collaboration:	Other
	Status of study:	One off data request
91.	**Project title:**	Parity information for selected anomalies for Berkshire
	Investigators:	Jenny Kurinczuk and Liz Ollerhead
	Collaboration:	Local
	Status of study:	One off data request
92.	**Project title:**	Neural tube defect and cardiac anomaly numbers
	Investigators:	Judith Rankin
	Collaboration:	Inter-register
	Status of study:	One off data request
93.	**Project title:**	Pulse OX trial – searching for cross border cardiac cases
	Investigators:	Ann Tonks
	Collaboration:	Inter-register
	Status of study:	One off data request
94.	**Project title:**	Spina bifida rates for statement in response to increase in Scotland
	Investigators:	Liz Draper
	Collaboration:	BINOCAR
	Status of study:	One off data request
95.	**Project title:**	FASP gastroschisis audit
	Investigators:	Anne Roberts
	Collaboration:	Local
	Status of study:	One off data request
96.	**Project title:**	Audit of outcome of antenatally diagnosed pulmonary lesions, ie congenital cystic adenomatoid malformation of lung (CCAM), pulmonary sequestration.

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	Investigators:	Peter Yeh
	Collaboration:	Local
	Status of study:	One off data request

97.	**Project title:**	Understanding the basis of abnormal haematopoeisis in babies with Down Syndrome
	Investigators:	Mark Anthony
	Collaboration:	Local
	Status of study:	One off data request

98.	**Project title:**	UKOSS/BAPS-CASS Study on CDH
	Investigators:	Marian Knight
	Collaboration:	Other
	Status of study:	One off data request

99.	**Project title:**	Consanguinity and Child Health - A Brief Health Needs Assessment (Oxfordshire PCT)
	Investigators:	Rosamund Southgate
	Collaboration:	Local
	Status of study:	One off data request

100.	**Project title:**	Consanguinity and Child Health - A Brief Health Needs Assessment (Buckinghamshire PCT)
	Investigators:	Lucy Jessop
	Collaboration:	Local
	Status of study:	One off data request

101.	**Project title:**	Audit of 11 conditions for the 20 week USS
	Investigators:	Jeanne Harris
	Collaboration:	Local
	Status of study:	One off data request

102.	**Project title:**	Downs babies case matching exercise for annual report
	Investigators:	Catryn Dixon, Alison Wainwright
	Collaboration:	Local
	Status of study:	One off data request

103.	**Project title:**	CCAM incidence to compare with Wessex region
	Investigators:	Diana Wellesley
	Collaboration:	Inter-register
	Status of study:	One off data request

104.	**Project title:**	Improving care for infants and their families before, during and after surgery.
	Investigators:	Jenny Kurinczuk
	Collaboration:	Local
	Status of study:	One off data request

105.	**Project title:**	FASP audit data supply
	Investigators:	Powatti Ramchand
	Collaboration:	Local
	Status of study:	One off data request

106.	**Project title:**	FASP audit data supply
	Investigators:	Annie Roberts
	Collaboration:	Local
	Status of study:	One off data request
107.	**Project title:**	Investigation into the genetic basis of renal tract anomalies - feasibility study
	Investigators:	Deirdre Cilliers
	Collaboration:	Local
	Status of study:	One off data request

Appendix 4

Publications to which CAROBB / OXCAR have contributed information

1. Best KE, Tennant PW, Addor MC, Bianchi F, Boyd P, Calzolari E, 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 Feb 1.

2. Bishop DV, Jacobs PA, Lachlan K, Wellesley D, Barnicoat A, Boyd PA, et al. Autism, language and communication in children with sex chromosome trisomies. Arch Dis Child. 2011 Oct;96(10):954-9.

3. Boyd PA, Haeusler M, Barisic I. EUROCAT Report 9: Surveillance of congenital anomalies in Europe 1980-2008. Birth Defects Res A Clin Mol Teratol. 2011 Mar;91 Suppl 1:S1.

4. Boyd PA, Haeusler M, Barisic I, Loane M, Garne E, Dolk H. Paper 1: The EUROCAT network--organization and processes. Birth Defects Res A Clin Mol Teratol. 2011 Mar;91 Suppl 1:S2-15.

5. Boyd PA, Loane M, Garne E, Khoshnood B, Dolk H. Sex chromosome trisomies in Europe: prevalence, prenatal detection and outcome of pregnancy. European Journal of Human Genetics. 2011 Feb;19(2):231-4.

6. Boyd PA, Tonks AM, Rankin J, Rounding C, Wellesley D, Draper ES. Monitoring the prenatal detection of structural fetal congenital anomalies in England and Wales: register-based study. Journal of Medical Screening. 2011;18(1):2-7.

7. Dolk H, Loane M, Garne E. Congenital Heart Defects in Europe: Prevalence and Perinatal Mortality, 2000 to 2005. Circulation. 2011 Mar 1;123(8):841-9.

8. Garne E, Dolk H, Loane M, Wellesley D, Barisic I, Calzolari E, et al. Paper 5: Surveillance of multiple congenital anomalies: implementation of a computer algorithm in European registers for classification of cases. Birth Defects Res A Clin Mol Teratol. 2011 Mar;91 Suppl 1:S44-50.

9. Greenlees R, Neville A, Addor MC, Amar E, Arriola L, Bakker M, et al. Paper 6: EUROCAT member registries: organization and activities. Birth Defects Res A Clin Mol Teratol. 2011 Mar;91 Suppl 1:S51-S100.

10. IPDTOC Working Group. Prevalence at Birth of Cleft Lip With or Without Cleft Palate: Data From the International Perinatal Database of Typical Oral Clefts (IPDTOC). The Cleft Palate-Craniofacial Journal. 2011;48(1):66-81.

11. Khoshnood B, Greenlees R, Loane M, Dolk H. Paper 2: EUROCAT public health indicators for congenital anomalies in Europe. Birth Defects Res A Clin Mol Teratol. 2011 Mar;91 Suppl 1:S16-22.

12. Loane M, Dolk H, Garne E, Greenlees R. Paper 3: EUROCAT data quality indicators for population-based registries of congenital anomalies. Birth Defects Res A Clin Mol Teratol. 2011 Mar;91 Suppl 1:S23-30.

13. Loane M, Dolk H, Kelly A, Teljeur C, Greenlees R, Densem J. Paper 4: EUROCAT statistical monitoring: identification and investigation of ten year trends of congenital anomalies in Europe. Birth Defects Res A Clin Mol Teratol. 2011 Mar;91 Suppl

1:S31-43.

14. Dolk H, Armstrong B, Lachowycz K, Vrijheid M, Rankin J, Abramksy L, et al. Ambient air pollution and risk of congenital anomalies in England, 1991-99. Occupational and Environmental Medicine. 2010 2010;67(4):223-7.

15. Garne E, Dolk H, Loane M, Boyd PA. EUROCAT website data on prenatal detection rates of congenital anomalies. Journal of Medical Screening. 2010;17(2):97-8.

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17. Garne E, Loane M, Addor MC, Boyd PA, Barisic I, Dolk H. Congenital hydrocephalus - prevalence, prenatal diagnosis and outcome of pregnancy in four European regions. European Journal of Paediatric Neurology. 2010;14:150-5.

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hernia:prenatal diagnosis, outcome and continuing morbidity in survivors. Br J Obstet Gynaecol. 1999;106:1192-96.

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Data Protection and handling requests for data

5a NIGB approval documentation

5b MREC approval documentation

5c Application form and guidelines for use of CAROBB data

Appendix 5

National Information Governance Board (NIGB) approval for CAROBB (as part of BINOCAR) to collect identifiable information without explicit consent from individuals registered.

Application Number	0011	
PIAG Reference	PIAG 2-08(e)/2002	
Other PIAG Refs		
Application Title	Congenital Anomalies Register (BINOCAR)	
Application Summary	To provide continuous epidemiological monitoring of the frequency, nature, cause and outcomes of congenital anomalies by means of national, regional and disease specific registers of congenital anomalies.**Dec 08 Application extended to contain address info at conception**	
Applicant Organisation Name	British Isles Network of Congenital Anomalies Register (BINOCAR)	
Contact Name	Elizabeth S Draper,Chair of BINOCAR	
Address	Department of Health Sciences, University of Leicester	
	22-28 Princess Road West	
	Leicester	
Postcode	LE1 6TP	
Telephone	0116 252 3210	
Fax		
Email	jlsb1@leicester.ac.uk	
Medical Purposes	Y	the surveillance and analysis of health and disease;
	□	the monitoring and audit of health and health related care provision and outcomes where such provision has been made;
	□	the planning and administration of the provision made for health and health related care;
	□	medical research approved by research ethics committees;
	□	the provision of information about individuals who have suffered from a particular disease or condition
Cohort/Population	UK-wide: patients with congenital anomalies	
Description of confidential patient information used	Mother's name, address, postcode, hospital number, NHS number, date of birth. Baby's name, address, postcode, hospital number, NHS number, date of birth, date of death. Address at conception.	
S60 Class(es)	□	Specific Support
	Y	Class I - making the person less readily identifiable
	Y	Class II - present or past geographical locations of patients
	Y	Class III - to identify and contact patients to obtain consent
	Y	Class IV - linking multiple sources;validating quality and completeness; avoiding error
	Y	Class V - audit, monitoring, & analysis of healthcare provision
	Y	Class VI - granting of access to data for purposes I-V
NHS Sponsor		
Status	Approved	
Date Applied		
Date Approved	20/06/02	
Date S60 Granted	20/06/02	
Expiry Date		
Next Review Date	05/08/2011	
Details of Approval	NIGB gave Section 60 support for the BINOCAR application.	
Notes		

Appendix 5b

NHS
National Research Ethics Service

Trent Research Ethics Committee
Research Ethics Office
Derwent Shared Services
Laurie House
Colyear Street
Derby
DE1 1LJ

Telephone: 01332 868765
Facsimile: 01332 868930

11 October 2009

Professor Elizabeth Draper
Dept of Health Sciences
22-28 Princess Road West
Leicester
LE1 6TP

Dear Professor Draper

Title of the Database: **British Isles Network of Congenital Anomaly Registers (BINOCAR)**
REC reference: **09/H0405/48**

The Research Ethics Committee reviewed the above application at the meeting held on 1 October 2009. Thank you for attending to discuss the application.

Ethical opinion

The members of the Committee present gave a favourable ethical opinion of the above research database on the basis described in the application form and supporting documentation.

Duration of ethical opinion

The favourable opinion is given for a period of five years from the date of this letter and provided that you comply with the conditions set out in the attached document. You are advised to study the conditions carefully. The opinion may be renewed for a further period of up to five years on receipt of a fresh application. It is suggested that the fresh application is made 3-6 months before the 5 years expires, to ensure continuous approval for the research database.

Approved documents

The documents reviewed and approved at the meeting were:

Document	*Version*	*Date*
Covering Letter		14 August 2009
REC application	IRAS Research Database Form V 2.3 (lock code 25660/56406/9/606)	19 August 2009
Participant Information Sheet	V 1.1	12 August 2009
Protocol	V 2.0	12 August 2009

Appendix 5b

Research governance

Under the Research Governance Framework (RGF), there is no requirement for NHS research permission for the establishment of research databases in the NHS. Applications to NHS R&D offices through IRAS are not required as all NHS organisations are expected to have included management review in the process of establishing the database.

Research permission is also not required by collaborators at data collection centres (DCCs) who provide data under the terms of a supply agreement between the organisation and the database. DCCs are not research sites for the purposes of the RGF.

Database managers are advised to provide R&D offices at all DCCs with a copy of the REC application for information, together with a copy of the favourable opinion letter when available. All DCCs should be listed in Part C of the REC application.

NHS researchers undertaking specific research projects using data supplied by a database must apply for permission to R&D offices at all organisations where the research is conducted, whether or not the database has ethical approval.

Site-specific assessment (SSA) is not a requirement for ethical review of research databases. There is no need to inform Local Research Ethics Committees.

Membership of the Committee

The members of the Ethics Committee who were present at the meeting are listed on the attached sheet.

Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees (July 2001) and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

After ethical review

Now that you have completed the application process please visit the National Research Ethics Service website > After Review

Here you will find links to the following:

a) Providing feedback. You are invited to give your view of the service that you have received from the National Research Ethics Service and the application procedure. If you wish to make your views known please use the feedback form available on the website.
b) Annual Reports. Please refer to the attached conditions of approval.
c) Amendments. Please refer to the attached conditions of approval.

Continued/

We would also like to inform you that we consult regularly with stakeholders to improve our service. If you would like to join our Reference Group please email referencegroup@nres.npsa.nhs.uk

09/H0405/48	Please quote this number on all correspondence

Yours sincerely

Dr Ian Gaywood
Chair

E-mail: jenny.hancock@derwentsharedservices.nhs.uk

Enclosures: *List of names and professions of members who were present at the meeting and those who submitted written comments*

Approval conditions

Appendix 5c

Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire

DATA REQUEST FORM

- This form is intended for requests for data for research purposes.
- Please read the CAROBB Guidelines and the notes on page 2 of this form before you sign.
- All requests will be approved by CAROBB Management Committee.
- Please complete, then **email and post a hard copy** (with signature and supporting documentation eg protocol) to Cath Rounding (CAROBB Co-ordinator) at the address at the bottom of this sheet.
- Please include any details of ethical approvals sought / granted.

Requester details	
Name:	
Job Title/Position:	
Organisation:	
Address:	
Contact phone number:	
Email address:	
Lead Clinician/Supervisor:	

Requester agreement	
Details of funding and source for project	
Name of person responsible for data security	
Request details	
Name of Project	
What question do you wish to answer?	
Intended use of information (e.g. Background, intended presentation/meeting/report)	

CAROBB, NPEU, University of Oxford, Old Road Campus, Headington, Oxford OX3 7LF
Direct: 01865 289721, Confidential fax: 01865 289720, E-mail: catherine.rounding@npeu.ox.ac.uk

Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire

DATA REQUEST FORM

Main outcome measures		
Case definition: (e.g. pre / postnatal diagnosis, live / stillbirths / TOPs.)		
Population: (e.g. CAROBB, Oxfordshire only)		
Time period (births):	By EDD or by Date of Birth? from to:	
Justification for identifiable data		
Do you plan to seek ethical approval / R&D approval for this study? (Please give details if yes)		
Signature:		Date:
Date required by:		

Please tick to confirm that you agree to the following:

- ☐ To supply CAROBB with a 6 monthly update report.
- ☐ On completion of the project all individual records will be destroyed and a CAROBB data destruction form completed and returned (requests for individual records only).
- ☐ Any publications/reports arising from the use of data supplied must include a standard acknowledgement paragraph (CAROBB will supply content).
- ☐ Any publications arising from the use of data supplied must be sent to Register Leads for approval while at draft stage. The register is also obliged to send the draft to the register funding body for approval.
- ☐ I have read and agree to the CAROBB Guidelines

We are keen for the CAROBB information to be used for research purposes and will do our best to help with any requests for data. Please do not hesitate to contact us with any queries.

CAROBB, NPEU, University of Oxford, Old Road Campus, Headington, Oxford OX3 7LF
Direct: 01865 289721, Confidential fax: 01865 289720, E-mail: catherine.rounding@npeu.ox.ac.uk

Appendix 5c

GUIDELINES for users CAROBB

Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire

CAROBB (Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire) was awarded funding by the Department of Health in 2003 to establish a database of information on babies born with suspected or confirmed congenital anomalies for the three counties. Prior to 2003, the register was known as OXCAR and included cases seen at the John Radcliffe Hospital since 1991.

The principal objectives of CAROBB are:

- Provide data for research on the aetiology and natural history of particular malformations to enable better advice based on accurate information to be given to parents
- Enable the evaluation and monitoring of new invasive and non invasive prenatal tests.
- Evaluate new prenatal screening programmes and to provide data for health care policies and planning
- Provide data to investigate clusters of abnormalities and putative teratogens by the monitoring of incidence over time and in population trends such as maternal age, ethnicity, and health inequalities.

CAROBB can be used as a basis for other studies and there are increasing numbers of requests for access to the data for research purposes. The Management Group wishes to encourage the use of the register in this way and the following guidelines have been drawn up to help potential register users. CAROBB conforms to the Data Protection Act 1998 and the Health and Social Care Act 2001.

Please feel free to contact the Register Co-ordinator for a discussion of your proposal at an early stage. It is important to be clear about what information you wish to collect and what information you will be able to obtain through the register.

1. All requests for access to CAROBB data should be made through the research co-ordinator using the accompanying form.

2. The request should be accompanied by a study protocol. The protocol must be approved by CAROBB. Approval by an ethics committee will not guarantee approval by CAROBB. Any amendments required by an ethics committee must be approved by CAROBB before data will be released.

3. If appropriate, the researcher will be responsible for obtaining approval from Ethics Committees in the areas in which the cases live. A copy of the approval must be supplied to the register co-ordinator before data will be released for the study.

4. Researchers are expected to seek peer review of the proposed study.

5. Researchers will need to seek the permission of the parent/child's general practitioner prior to contacting parents and children. If necessary, permission must

also be sought from the appropriate consultant for access to hospital notes.

6. If the researcher has little or no previous experience of research the Management Group will require a written assurance from a supervisor that the work will be carried out and completed satisfactorily.

7. It is the responsibility of the researcher to apply for funds to carry out the proposed study. A small administrative charge may be made to cover the cost of accessing cases from CAROBB.

8. Data supplied by CAROBB must not be passed to a third party, nor should it be re-used for later study without applying to CAROBB for permission. Personal data must not be uploaded to a researchers home computer. Researchers are expected to deposit datasets which have been derived from the original data, with suitable documentation, in the CAROBB database.

9. In compliance with the Data Protection Act, 1998, to keep the database as accurate as possible, researchers will be expected to inform CAROBB of changes to subjects details during the course of the study.

10. The Management Group will request a short progress report at intervals during the course of the study and evidence of the final results in the form of a report or paper. Any change in contact addresses or personnel working on the project should be notified to the Management Group.

11. The Management Group would like to see an advanced draft of any publication, or abstract submitted for a meeting, in which CAROBB data have been used. Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire should be acknowledged in any publication or presentation, arising from CAROBB data, using the sentence "The Management Group of Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire approved the release of register data for this study. CAROBB is funded by the Department of Health."

12. On completion of the analysis and after copy datasets have been supplied to CAROBB, ALL PERSONAL IDENTIFIABLE INFORMATION MUST BE DESTROYED, in accordance with any requirements of the ethics approval for the study. If you are unsure on this point, contact CAROBB for clarification.

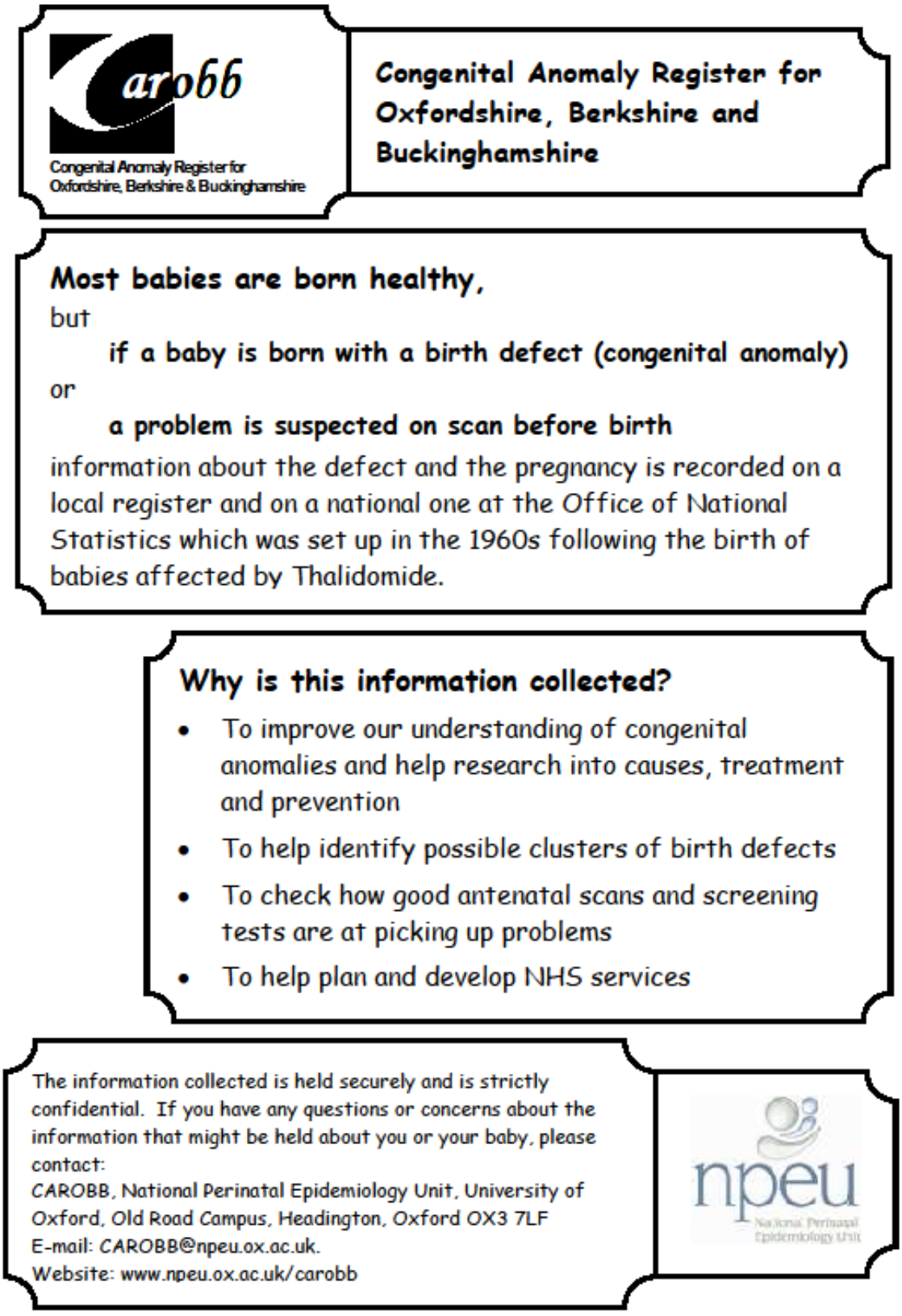
Please complete the application form enclosed and return to the CAROBB office.

Publicity

6a Poster for clinic waiting rooms

6b Leaflet for clinic waiting rooms

Appendix 6a


carobb
Congenital Anomaly Register for Oxfordshire, Berkshire & Buckinghamshire
Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire
Most babies are born healthy,
but
if a baby is born with a birth defect (congenital anomaly)
or
a problem is suspected on scan before birth
information about the defect and the pregnancy is recorded on a local register and on a national one at the Office of National Statistics which was set up in the 1960s following the birth of babies affected by Thalidomide.
Why is this information collected?
To improve our understanding of congenital anomalies and help research into causes, treatment and prevention
To help identify possible clusters of birth defects
To check how good antenatal scans and screening tests are at picking up problems
To help plan and develop NHS services
The information collected is held securely and is strictly confidential. If you have any questions or concerns about the information that might be held about you or your baby, please contact:
CAROBB, National Perinatal Epidemiology Unit, University of Oxford, Old Road Campus, Headington, Oxford OX3 7LF
E-mail: CAROBB@npeu.ox.ac.uk.
Website: www.npeu.ox.ac.uk/carobb
npeu
National Perinatal Epidemiology Unit

How is information collected?

A member of staff from the hospital who treats you or your baby, completes a notification to the register when the anomaly is identified. The register often receives several notifications from different departments about the same baby. Any information reported in the early stages can be improved or confirmed later by these multiple notifications.

Names and postcodes are included so that information can be updated on the correct case and the same baby is not counted several times.

Information is collected on paper and stored electronically on a computer. This information is held securely by CAROBB, which is based at The National Perinatal Epidemiology Unit, in Oxford.

Does my name or my baby's name have to go on the Register?

We hope everyone will want to be included on the Register, to help us plan and improve services for future mothers and babies. However, your details can be removed at any time.

Will the database be secure and confidential?

The information recorded on the Register about you or your baby is confidential. It is held in a responsible way which respects the rights and privacy of individuals.

The Register follows a strict policy on security and confidentiality. This policy is available to the public. The register conforms to the requirements of legislation on data protection.

How can I find out more about CAROBB?

If you have any questions or concerns regarding the information that could be held on you or your baby, please contact the registry:

CAROBB
National Perinatal Epidemiology Unit
University of Oxford
Old Road Campus
Headington
Oxford OX3 7LF

Tel: 01865 289721
Fax: 01865 289720
E-mail: carobb@npeu.ox.ac.uk
Website: www.npeu.ox.ac.uk/carobb/

CAROBB and The National Perinatal Epidemiology Unit are funded by the Department of Health

Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire

Information for parents

Every parent hopes that their baby will be healthy and most babies are.

However, a few babies do have problems (abnormalities) such as cleft palate, spina bifida, or Down's syndrome. These are sometimes called congenital anomalies or congenital malformations.

Some congenital anomalies are detected during pregnancy, some are found at birth, while others become apparent only as a baby grows older.

Why is information collected about babies with congenital anomalies?

CAROBB collects information:

- To increase our understanding of congenital anomalies and help research into their causes, treatment and prevention.
- To monitor how good antenatal screening tests (serum screening and ultrasound scans) are at picking-up problems.
- To look at trends - for example changes in the number of babies born with congenital anomalies, or changes in the pattern of where they are born.
- To give health professionals information to help them advise families about their chances of having a baby with a congenital anomaly.
- To help plan and develop NHS services.

What is CAROBB?

***CAROBB* (Congenital Anomaly Register for Oxfordshire, Berkshire and Buckinghamshire) is a database of information on babies born with suspected or confirmed congenital anomalies.**

What information is collected?

Information held by the register includes:

- Descriptions of each anomaly.
- Details and results of any investigations carried out during pregnancy (for example, the results of any ultrasound scans).
- Details about mother and baby.

Who sees the information?

There are very strict regulations controlling access to personal information - that is names and addresses. This information will only be available to members of hospital staff treating you or your baby, and to those who work on CAROBB.

Information is also sent to the National Congenital Anomaly Surveillance System, which collects information for the whole country. When this happens only the first three letters of the baby's name are sent.

Information that is used by researchers or published in reports does not contain anything to identify either mother or baby, such as names and addresses.

Can I see the records on the Register?

Yes - you have the right to request a copy of the information held on you or your baby.

To do this, please make your wishes known to a member of your healthcare team or contact CAROBB by telephone or e-mail