

# caris review 2013

including data 1998 – 2012

Welcome to the new CARIS leaflet which replaces the annual report.

2013 is a big year for CARIS. We are now 15 years old and have collected information on over 25,000 babies on a birth population of over half a million.

Thank you for your contributions and support over the years.

CARIS aims to provide reliable data on congenital anomalies in Wales which can be used to assess patterns of anomalies, including possible clusters and their causes and to inform the work of health services, including antenatal screening.

## Patterns of anomalies

During 1998-2012 there were 25,650 cases of congenital anomalies reported to CARIS (22,039 live born) out of 501,894 total (live and still) births in Wales.

- The gross rate of congenital anomalies reported is 5.1%\*
- The rate of congenital anomalies in live born babies is 4.4%
- 85.9% of cases are live born and 96.7% of these survive to the end of the first year of life. Survival is reduced with increasing complexity of anomalies.
- 57.9% of cases involve a single congenital anomaly. 10.6% of cases are associated with an underlying chromosomal disorder.

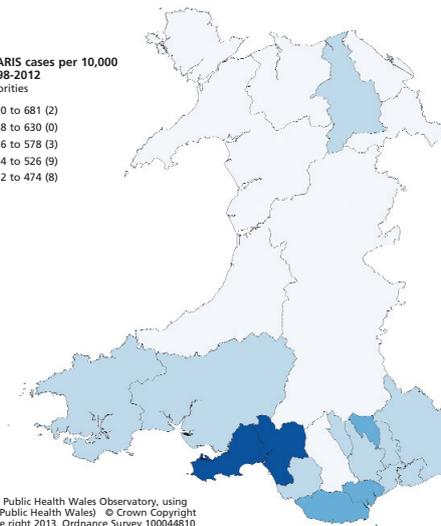
\* Gross rate is the total number of cases of anomaly (regardless of whether the pregnancy ended in miscarriage, termination of pregnancy, live birth or stillbirth) divided by the total number of live and still births.



## Gross rate of CARIS cases, 1998-2012

Rate of CARIS cases per 10,000 births, 1998-2012  
Local Authorities

630 to 681 (2)
578 to 630 (0)
526 to 578 (3)
474 to 526 (9)
422 to 474 (8)

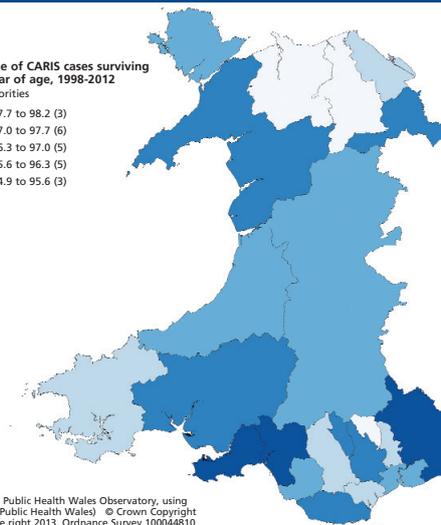


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## Percentage of CARIS cases surviving to one year of age, 1998-2012

Percentage of CARIS cases surviving to one year of age, 1998-2012  
Local Authorities

97.7 to 98.2 (3)
97.0 to 97.7 (6)
96.3 to 97.0 (5)
95.6 to 96.3 (5)
94.9 to 95.6 (3)



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The gross rate of congenital anomalies ranges from 4.2% in Flintshire to 6.8% in Swansea. Rates are thought to be affected by variations in reporting. Good reporting is likely to identify less severe cases and this is reflected, to some degree, in the survival patterns.

## Working with services

**Newborn bloodspot screening** detects rare but serious diseases that respond to early intervention. The screening test is undertaken ideally on day five of life and is part of routine post natal care. In Wales, screening was introduced for medium-chain acyl-CoA dehydrogenase deficiency from June 2012 and sickle cell disorder from June 2013.

We focus on the metabolic conditions screened after birth in this year's CARIS meetings. More detail can be found on our website and at [www.newbornbloodspotscreeningwales.nhs.uk](http://www.newbornbloodspotscreeningwales.nhs.uk)



### Conditions detectable by newborn bloodspot screening, recorded by CARIS, numbers and birth ratios, 1998-2012

	Number	Wales	Orphanet estimate*
Congenital hypothyroidism	307	1: 1,600	1: 2,000 to 1: 4,000
Cystic fibrosis	223	1: 2,300	1: 8,000 to 1: 10,000
Phenylketonuria (PKU)	46	1: 10,700	1: 15,000
Sickle cell disorders†	17	n/a	1: 2,000 to 1: 10,000
Medium-chain acyl-CoA dehydrogenase deficiency (MCADD) †	4	n/a	1: 4,900 to 1: 17,000

\* Orphanet estimates or research to which Orphanet links; based on dates and populations which are not always comparable.

Source: [www.orphanet.net](http://www.orphanet.net)

† Birth ratio not displayed as conditions were not part of the newborn bloodspot screening programme in Wales for most of the period.

**Antenatal detection** is improving in Wales. CARIS data shows this improvement, particularly for cardiac anomalies. 56.1% of severe cardiac defects were detected antenatally during 2010-2012. Wales now has the best antenatal detection of heart defects in the UK. This improvement was recognised with an NHS Wales Award during 2013.

## Survival of babies with certain congenital anomalies to age five (1998-2006)

With 15 years of data, CARIS can look back at survival of babies with anomalies over an extended period. Over 90% of children born with cystic fibrosis, spina bifida, Turner syndrome and gastroschisis in Wales survived past their fifth birthday. Until the 1990s, babies born with hypoplastic left heart syndrome had little hope of survival but with the development of new surgical procedures 43% of babies now survive past their fifth birthday.

Total cases*	Liveborn cases	Prevalence at 5th birthday**
140	135 (96%)	4.4
236	43 (18%)	1.4
130	37 (28%)	1.2
181	161 (89%)	5.1
629	290 (46%)	8.5
118	93 (79%)	2.6
283	143 (51%)	3.9
32	31 (97%)	0.7
119	83 (70%)	2.0
102	46 (45%)	0.7

- % liveborn babies who died before 1st birthday
- % liveborn babies who died between 1st and 5th birthday
- % liveborn babies who survived to age five years

Cystic fibrosis	2	98
Spina bifida	5	95
Turner syndrome	5	95
Gastroschisis	6	94
Down syndrome	9	88
Transposition	15	84
Hydrocephaly	15	80
TAPVC	23	71
CDH	25	73
HLHS	48	43

Data source: CARIS (PHW) & MYE (ONS) \* where year of end of pregnancy between 1998 and 2006. Includes fetal losses, termination of pregnancies, stillbirths and livebirths. Please note due to rounding the percentages totals do not always equal 100.

\*\* rate per 10,000 population (5 year olds in period 2003-2011). TAPVC = Total anomalous pulmonary venus connection

HLHS = Hypoplastic left heart syndrome CDH = Congenital diaphragmatic hernia Transposition = Transposition of the great vessels

## Data quality

CARIS is part of the EUROCAT network. Wales performs consistently well in data quality compared with other registers. For more information go to: [www.eurocat-network.eu/aboutus/datacollection/dataquality/dataqualityindicators](http://www.eurocat-network.eu/aboutus/datacollection/dataquality/dataqualityindicators)

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The CARIS team (l/r) Helen Jenkins, David Tucker, Margery Morgan, Judith Greenacre, Val Vye and Vivian Morgan (not pictured).