

# caris review 2015

including data 1998 – 2014

These are the key findings from the annual update of the congenital anomaly register. This now covers the years 1998 to 2014 and contains information about nearly 30,000 cases. More information can be found on the CARIS website at:

[www.wales.nhs.uk/caris](http://www.wales.nhs.uk/caris)

## CARIS provides reliable data on congenital anomalies in Wales to:

- Assess patterns of anomalies, including possible clusters and their causes
- Inform the work of other health services, including screening

Our thanks go to everyone involved in providing us with information. This has enabled us to build and maintain one of the most comprehensive national congenital anomaly databases in Europe.



## Patterns of anomalies

- Between 1998 and 2014, 29,642 cases of congenital anomaly were reported to CARIS, out of a total of 569,537 (live and still) births in Wales. This gives a gross rate<sup>1</sup> of 5.2%.
- 58% of those affected were male, and 40% female. 14 were described as intersex, and the remainder were unknown or not recorded, often because the pregnancies ended in a miscarriage or termination.
- 25,540<sup>2</sup> (86.2%) of the babies were live born and 96.9% of these (born between 1998 and 2013) lived until at least their first birthday. Survival was reduced where there were increasingly complex anomalies.
- 60.2% of cases were affected by a single congenital anomaly. 11.3% of the cases were associated with an underlying chromosomal disorder.
- The five most common groups of anomaly in descending order are circulatory, limb, musculoskeletal, urinary and digestive.

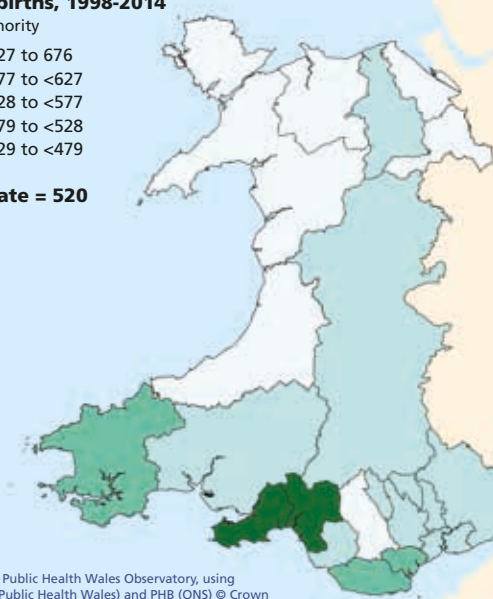
The variations in rates shown on both maps may be due to variation in reporting of anomalies.

## Gross rate of CARIS cases, 1998-2014

Rate of CARIS cases per 10,000 births, 1998-2014  
Local Authority

627 to 676
577 to <627
528 to <577
479 to <528
429 to <479

Wales rate = 520



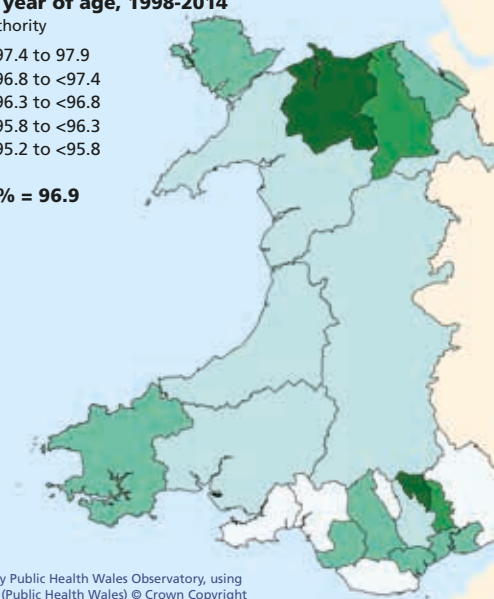
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## Percentage of CARIS cases surviving to one year of age, 1998-2014

Percentage of CARIS cases surviving to one year of age, 1998-2014  
Local Authority

97.4 to 97.9
96.8 to <97.4
96.3 to <96.8
95.8 to <96.3
95.2 to <95.8

Wales % = 96.9



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<sup>1</sup> Gross rate is the total number of cases of anomaly (whether the pregnancy ended in live birth, still birth, termination or miscarriage) divided by the total number of live and still births.

<sup>2</sup> This is the total number of babies with an anomaly born between 1998 and 2013. Any babies born during 2014 will not have reached their first birthday during 2014.

There are two areas of focus this year – congenital disorders of sex development and teenage pregnancies.

## Sex development disorders, Wales, 1998 -2014

Anomaly	Total cases	Average cases per year	Rate per 10,000 total births	% of cases liveborn
Hypospadias	1,702	100	29.9	99.6
Cryptorchidism (undescended testes)	1,382	81	24.3	99.8
Klinefelter's Syndrome	63	4	1.1	76.2
Congenital adrenal hyperplasia	46	3	0.8	97.8
Mixed gonadal dysgenesis	22	1	0.4	72.7
Micropenis	17	1	0.3	64.7
Androgen insensitivity	6	<1	0.1	83.3

Produced by Public Health Wales Observatory, using CARIS & PHB (ONS)

## Disorders of sex development – Wales 1998-2014

Most of these conditions are rare and most are not diagnosed antenatally. The two most common disorders – hypospadias and cryptorchidism may be diagnosed at birth but clinical reporting to CARIS is poor. Information about both disorders is often delayed until there is notification of surgery to correct the disorders. Other conditions are very rare as is shown in the table.

Parents with a baby with a disorder of sex development at birth will need guidance from an experienced clinician, usually a paediatric endocrinologist. Ideally their child will be seen by a specialist multidisciplinary team with experience of caring for children with these disorders. Members of the team should include specialists in surgery, urology, psychology/psychiatry, radiology and nursing. Wider access to specialists in genetics, gynaecology, plastic surgery, biochemistry and clinical ethics may also be necessary.



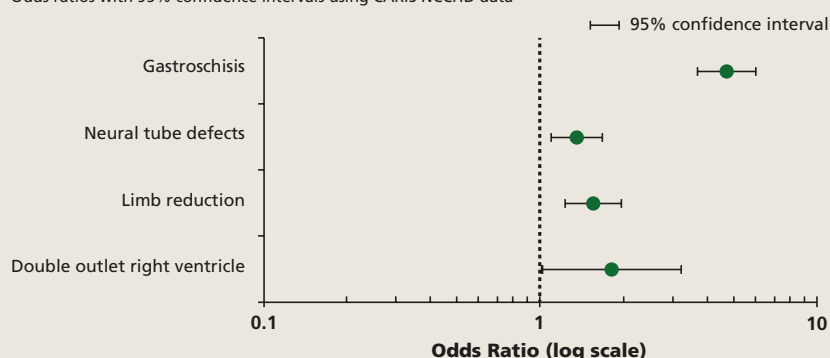
## Teenage pregnancy and congenital anomalies

Young women who become pregnant while they are teenagers (<20 years) are at increased risk of their baby developing a congenital anomaly. Although across the UK the teenage pregnancy rate is falling, births to UK women<sup>3</sup> aged under 20 are the 4th highest in Europe after Bulgaria, Romania and Slovakia. Within the UK the rate is consistently higher in Wales than in England: in 2013 the maternity rate for under 18s in Wales was 14.9/1,000 total births compared to the English rate of 11.9/1,000<sup>4</sup>.

Using CARIS data, a comparison was made between congenital anomalies in teenagers (<20 years) and women in the 20-34 year age group<sup>5</sup>.

### Increased risk of specific congenital anomalies in teenage pregnancies in Wales (1998 to 2012)

Odds ratios with 95% confidence intervals using CARIS NCCHD data



This study found significantly increased rates of neural tube defects, gastroschisis, limb reduction defects and double outlet right ventricle. Of these, gastroschisis has been well recognised in younger mothers. Research is planned to look at these conditions in terms of risk factors in teenage mothers.

The significantly raised numbers of neural tube defects in the teenage population suggests a value in targeting these youngsters in terms of planning pregnancy, taking preconception folic acid and eating a healthy diet.

<sup>3</sup> Eurostat data, compiled by the Office for National Statistics 'Live birth rate (per 1,000) to women aged 15-19 and 15-17 in EU28 countries, 2012' from <http://www.ons.gov.uk/ons/rel/vsob1/births-by-area-of-usual-residence-of-mother--england-and-wales/2012/sty-international-comparisons-of-teenage-pregnancy.html> accessed September 2015

<sup>4</sup> Office for National Statistics 'Conception statistics, England and Wales, 2013'

<sup>5</sup> Teenage Pregnancy Anomaly Rates in Wales, UK – Which systems have Increased Risk? Hodge et al, ICBDsr 2015 [centre@icbdsr.org](http://centre@icbdsr.org)

## 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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