

caris review 2016

data from 1998 to 2015

This short report contains information about the CARIS register to the end of 2015, which now contains details of 31,123 cases. More detailed information and data tables can be found at:

www.wales.nhs.uk/caris

CARIS background

CARIS was established in 1998 with the objective of assessing patterns of anomalies in Wales, including possible clusters and their causes. For example it is known that Wales has the second highest rate of neural tube defect in Europe, and there is ongoing work looking into the causes of this, and to see if there is further potential for prevention. Information from CARIS informs planning of wider health services, including screening services. CARIS could not operate without the hard work and good will of all who provide us with the information to build a comprehensive congenital anomaly database.



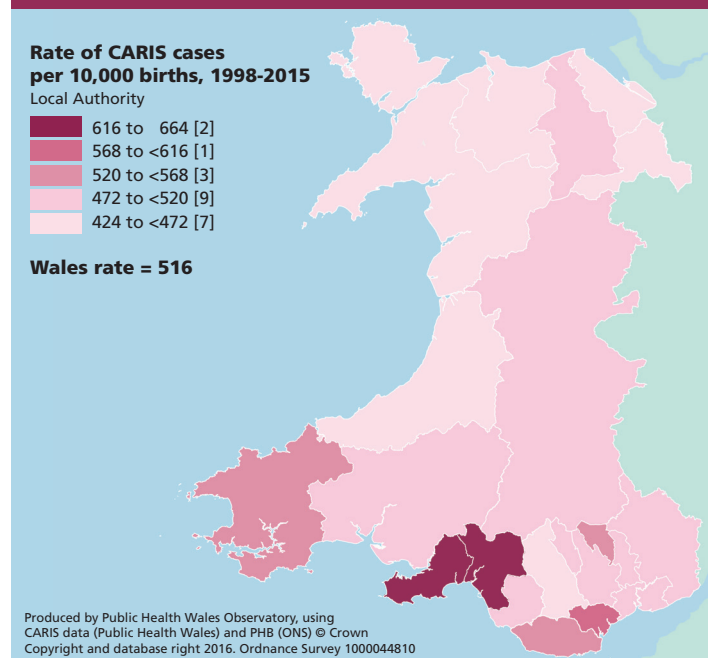
Patterns of anomalies

- 31,123 cases of congenital anomaly were reported to CARIS between 1998 and 2015. There were a total of 602,972 (live and still) births in Wales during this time. This gives a gross¹ rate of 5.2%, unchanged since last year's review.
- 26,816 were live born (86.2%) of whom 96.9% survived until their first birthday (babies born between 1998 and 2014). Survival was reduced where very complex anomalies were observed.
- Most (59.9%) of the cases had a single anomaly, and 11.6% had an underlying chromosomal disorder. The rate of chromosomal disorders is increasing each year. This may relate to increased maternal age and the availability of genetic screening
- 40% of those affected were female, 58% were male, and 14 were described as intersex. The remainder were unknown or not recorded, often because the pregnancies ended in termination or miscarriage.
- The five most common groups of anomaly in descending order were circulatory, limb, musculoskeletal, urinary and digestive. This is unchanged since last year's review.

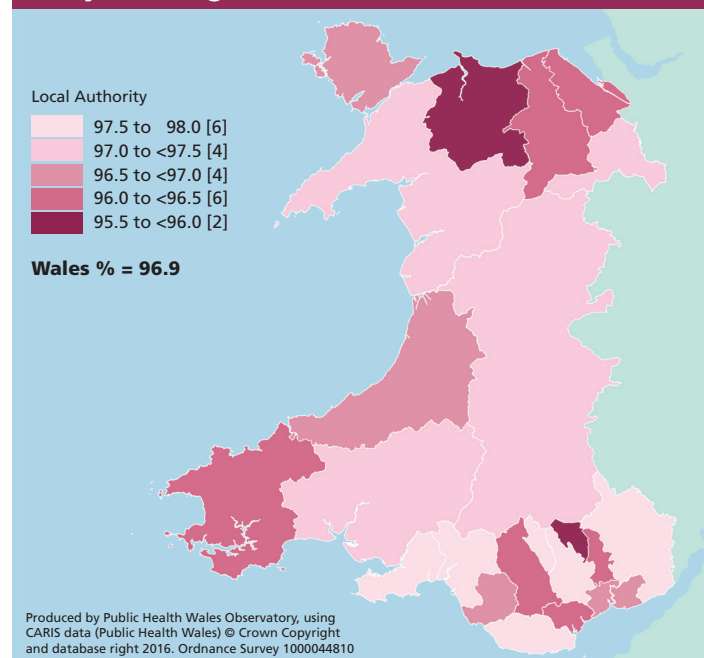
Across Wales, as can be seen from Map 1, there is variation in the rates. However this may well be due to variations in reporting.

¹ 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.

Map 1 gross rate of CARIS cases, 1998 – 2015



Map 2 percentage of CARIS cases surviving to one year of age, 1998 – 2015



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



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The two areas of focus for this year's review are infections in pregnancy and blood disorders.

Infections in pregnancy

Congenital infections have been in the news recently with the emergence of Zika virus in south America, thought to be linked to microcephaly², no cases have been reported in Wales, but surveillance for this is in place. There are a number of infections which, if contracted by a woman during or before pregnancy, can have serious consequences for the unborn child. These infections include cytomegalovirus (CMV), herpes, syphilis, toxoplasmosis, parvovirus, chicken pox and rubella. The complications caused by these infections vary in type and severity, and are affected by the gestational age when the infection is contracted.

The number of cases of congenital anomaly due to infection³ during pregnancy which are notified to CARIS is small – a total of 133 cases of congenital anomaly related to maternal infection during pregnancy between 1998 and 2015. This gives an average of seven cases per year, with a rate of 2.2 per 10,000 total births. Of these 133 cases, 73% were live born. Because of the small numbers no breakdown by specific infection is available. The most commonly reported infection is CMV, followed by parvovirus and toxoplasmosis.

Women are screened antenatally for some infections such as HIV, Hepatitis B and syphilis. Screening for rubella susceptibility stopped in October 2016, after guidance from the UK National

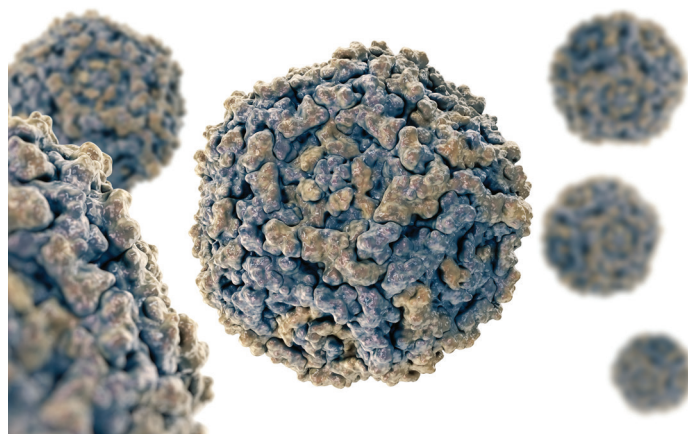


Image: Parvovirus

Screening Committee. No cases have been detected in Wales since CARIS was established. Nor have any cases of neonatal transmission of HIV been reported to CARIS. Guidance for health professionals who are concerned about infections and rashes in pregnancy has been updated and can be found here: <http://www.antenatalscreening.wales.nhs.uk/professional/document/283802>

² <http://www.wales.nhs.uk/sitesplus/888/page/84861>

³ Congenital Infections (A50*, P35* excluding P35.3 & P37* excluding P37.5

Blood disorders

There are a large number of congenital blood disorders, some more common than others. Blood disorders include conditions that affect both red and white blood cells, platelets, bone marrow, and the blood constituents involved in clotting or bleeding.

Cases with disorders of the blood and blood forming organs, rates per 10,000 total births and percentage of cases liveborn, Wales 1998-2015

Anomaly	Total cases	Average cases per year	Rate	% of cases liveborn
Hereditary Spherocytosis	68	4	1.1	100.0
Haemophilia A	39	2	0.6	100.0
Von-Willebrands Disease	34	2	0.6	100.0
Sickle-cell Anaemia	27	2	0.4	96.3
Thalassaemia	25	1	0.4	96.0

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

Information about the five most commonly occurring disorders reported to CARIS is displayed in the table, but even though they are the most common, numbers are still very small. There is under reporting of some conditions because they can be mild with few symptoms, or because some (e.g. haemochromatosis) only become apparent later in life, and are not always reported to CARIS. The blood spot test carried out on every newborn baby in Wales includes a test for sickle cell disease. More information about blood disorders will be available on the CARIS website.



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