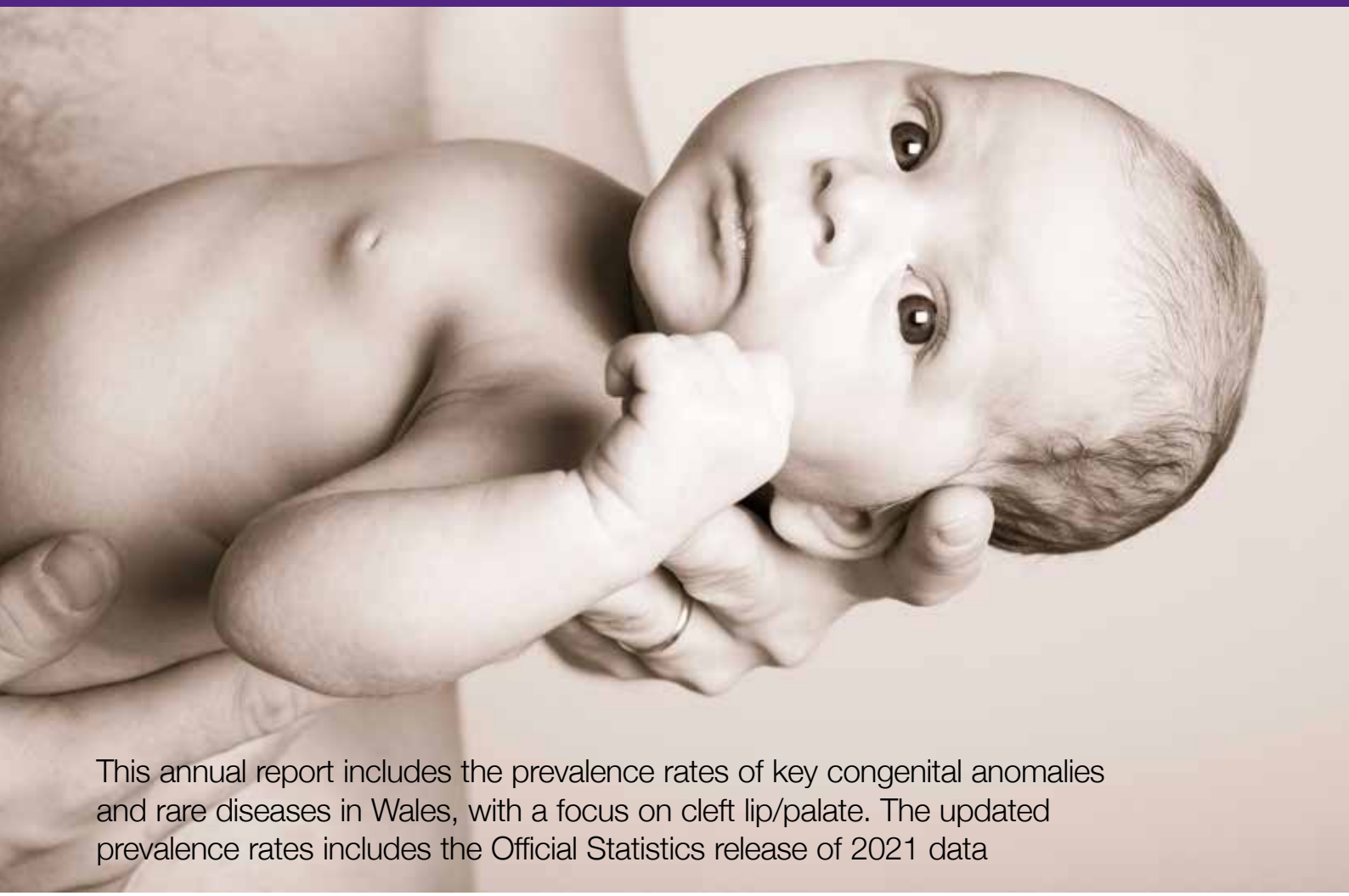
Congenital Anomaly Register & Information Service for Wales

caris review 2021

data from 1998 to 2021



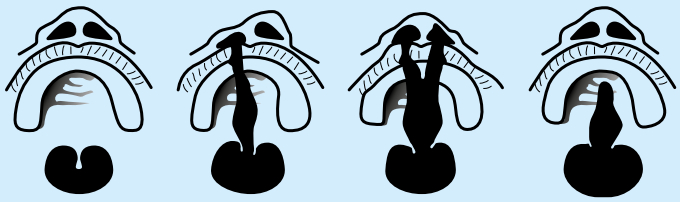
This annual report includes the prevalence rates of key congenital anomalies and rare diseases in Wales, with a focus on cleft lip/palate. The updated prevalence rates includes the Official Statistics release of 2021 data

Facial Clefting

This is the most common anomaly of the face. Cleft lip and cleft palate may occur in isolation or together and may be associated with other facial defects. A cleft lip results from failure of the median nasal swellings to fuse with the maxillary swellings and a cleft palate is a result from a failure of the roof of the mouth to join completely. This is usually in the second month of pregnancy.

Cleft lip with or without cleft palate occurs in about 2 in 1000 livebirths in Wales with higher levels in some racial groups. Boys are affected more often than girls and about 13% of cases are associated with other anomalies. Unilateral cleft lip is more common than bilateral, and the left side is more commonly affected than the right. Risk factors include genetic factors, smoking, excess alcohol intake and anticonvulsant medication. Maternal folic acid may reduce the risk.

FIGURE 1 Facial Clefting



Contact us

CARIS, Public Health Wales, Level 3 West Wing Block, Singleton Hospital, Sketty Lane, Swansea SA2 8QA
Tel: 01792 285241 (WHTN 01883 6122) Email: CARIS@wales.nhs.uk Caris.SafehavenMailbox@wales.nhs.uk

GIG CYMRU NHS WALES
Iechyd Cyhoeddus Cymru
Public Health Wales

Isolated cleft palate appears to be a separate entity. In Wales it occurs in up to 1 in 1000 livebirths with no racial preference. Girls are more frequently affected than boys and about 50% are associated with other anomalies, especially chromosomal defects. Risk factors include genetic factors, increased maternal age, smoking, alcohol and anticonvulsants. Folic acid does not reduce the risk.

Pierre Robin sequence is a condition with a cleft palate where the baby has a small lower jaw (micrognathia) and a backward-positioned tongue (glossoptosis).

Most antenatal diagnosis takes place at the 20-week scan. A cleft lip can show as a vertical hypoechoic (appearing black) area within the upper lip, usually to the left of the midline. A bilateral cleft lip and palate can show as an echodense mass in the centre of the upper lip. 3D scanning, colour flow Doppler and magnetic resonance can be of value particularly with an isolated cleft palate. After finding a cleft, a check for other anomalies is carried out because of the increased chance of associated defects (particularly chromosomal disorders and heart anomalies).

The best treatment involves a multidisciplinary approach which can include a paediatrician, plastic surgeon, dentist, ENT specialist, speech therapist, audiologist, genetic counsellor and social worker.

Surgery aims to correct a cleft lip and palate both functionally and cosmetically. The lip is usually repaired at 3-6 months of age. A cleft palate is repaired between 6 and 18 months of age.

FIGURE 2 Cleft lip/palate profile in Wales n=1802

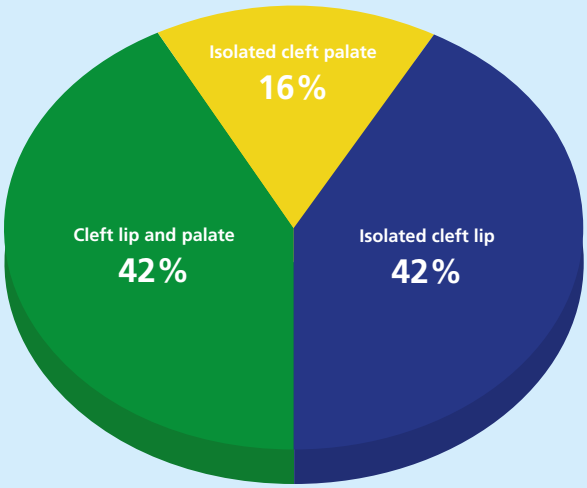
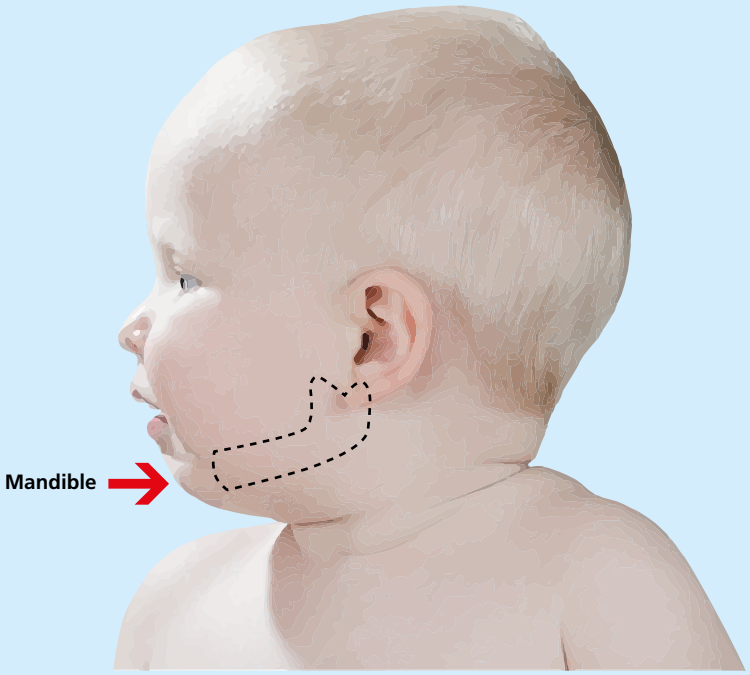


FIGURE 3 Pierre Robin sequence



Challenges for the Baby

- Feeding difficulties – mainly cleft palate, may need special bottles
- Ear infections and hearing loss – mainly in cleft palate, annual checks of middle ear for fluid required
- Speech challenges – worse in cleft palate with nasal speech and consonant difficulties
- Dental problems – some clefts affect the gum resulting in missing teeth and higher risk of caries

www.nhs.uk/conditions/cleft-lip-and-palate/treatment
CARIS review 2010 *phw.nhs.wales/services-and-teams/caris*
Bernheim N, Georges M, Malevez C, De Mey A, Mansbach A (2006) Embryology and epidemiology of cleft lip and palate. B-ENT Vol.2 Suppl. 4

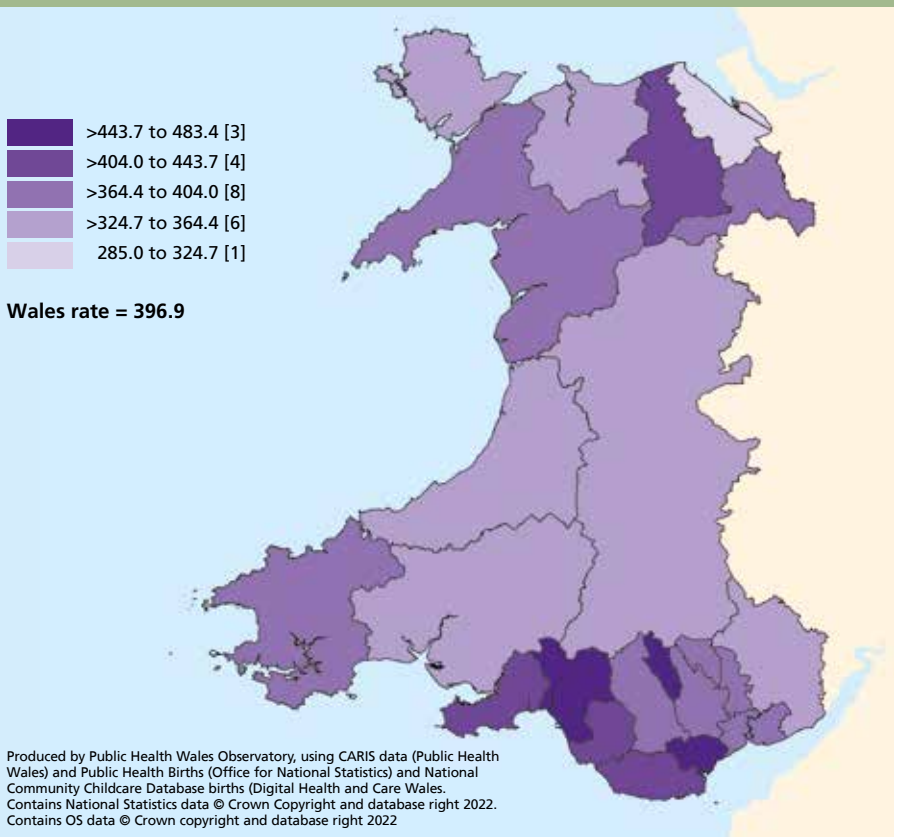
Update on Congenital Anomalies 2021

Since 1998 the CARIS team have registered 38,528 congenital anomaly cases in Wales. Of all live and still births in Wales, 4.9% are affected by a congenital anomaly. This proportion is slightly lower than the 5.1% reported at the 2018 update, however, it is acknowledged that the COVID pandemic may have impacted data collection. For example, during lockdowns routine surgical procedures may have been postponed thereby delaying reporting via this route.

The proportion of babies affected by a congenital anomaly that were liveborn was 85%, with 96.6% surviving to 1 year of age. Of those with a recorded gender, 59% were male. These proportions remain similar to those reported at the 2018 update.

The prevalence rates by local authority region remains largely unchanged from previous years and are summarised in Figure 4. Please note that this map only includes data collected in the past 10 years. In previous reports we have included maps containing data from the registry establishment in 1998. Therefore, please interpret with caution if comparing to previous years reports.

FIGURE 4 Rate of CARIS cases per 10,000 births 2012-2021, Wales



Produced by Public Health Wales Observatory, using CARIS data (Public Health Wales) and Public Health Births (Office for National Statistics) and National Community Childcare Database births (Digital Health and Care Wales).
Contains National Statistics data © Crown Copyright and database right 2022.
Contains OS data © Crown copyright and database right 2022

A singular anomaly was reported in 57.7% of cases and 13.9% had an underlying chromosomal disorder. The five largest groups of anomalies were circulatory, limbs, musculoskeletal, genetic/multi-site disorder and urinary. The rates for these groups were largely unchanged from the previous 2018 update. However, digestive disorders and genital disorders have dropped out of the largest five groups since the 2018 update. For digestive disorders the current rate is 64.3, down from 66.1 per 100,000 births in 2018. For genital disorders the current rate is 59.3, down from 61 per 100,000 births in 2018. These group specific rate reductions may be related to reporting delays due to the pandemic impact on, for example, elective surgery.

In addition to the headline summary provided in this report, the data tables and outputs that form our official statistics release relating to childhood rare diseases and antenatal detection rates are available here (INSERT WEBSITE LINK).

Research carried out by the CARIS team

Ten-Year Survival of Children With Congenital Anomalies: A European Cohort Study | Pediatrics | American Academy of Pediatrics (aap.org)

Epidemiology of Pierre-Robin sequence in Europe: A population-based EUROCAT study - Santoro - 2021 - Paediatric and Perinatal Epidemiology - Wiley Online Library

Analysis of early neonatal case fatality rate among newborns with congenital hydrocephalus, a 2000-2014 multi-country registry-based study - PubMed (nih.gov)

Behçet's disease in Wales: an epidemiological description of national surveillance data | Orphanet Journal of Rare Diseases | Full Text (biomedcentral.com)

Frontiers | The Improved Prognosis of Hypoplastic Left Heart: A Population-Based Register Study of 343 Cases in England and Wales (frontiersin.org)

Frontiers | Prevention of Neural Tube Defects in Europe: A Public Health Failure (frontiersin.org)

A Multicountry Analysis of Prevalence and Mortality among Neonates and Children with Bladder Exstrophy - PubMed (nih.gov)

A list of CARIS presentations can be found here Publications and Presentations - Public Health Wales (nhs.wales)

Authors

David Tucker – CARIS Manager, Public Health Wales
Margery Morgan – Lead Clinician CARIS, Obstetrician, Singleton Hospital Swansea
Llion Davies – Public Health Consultant, Public Health Wales
Samantha Fisher – Senior Registration & Analysis Officer CARIS, Public Health Wales

GIG CYMRU NHS WALES
Iechyd Cyhoeddus Cymru
Public Health Wales