Approximately 1,400 new cases are reported annually to CARIS. By the end of 2016, 32,580 cases had been recorded on the CARIS database since 1998. The rate of congenital anomalies is 5.1% and most cases (59.4%) are registered with a single anomaly. 86% of cases are born alive, giving a 4.4% live-born rate of babies with congenital anomalies. 96.9% of the babies who are live-born survive to their first birthday.

Of the cases where the sex is known, 59.1% are male and 40.9% are female. There are 804 cases where the sex is not recorded or not known (mostly due to termination or miscarriage) and 13 cases recorded as intersex. From the map it appears that there are more cases in the Swansea area than elsewhere, but this probably reflects better recording, as the CARIS office is situated in Singleton Hospital, Swansea.

A significant development during 2017 is that the CARIS registration team was given access to Badgernet – a software system which records all neonatal admissions in Wales. This means our case recording will be more comprehensive in the future.

A second development was success in a bid for participation in EUROlinkCAT, a European study providing information on mortality and outcomes for children born with congenital anomalies.

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* The 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.
Pre-gestational diabetes and congenital anomalies

Work has been undertaken reviewing congenital anomalies in women with pre-gestational type 1 and 2 diabetes. The rate of type 2 diabetes is increasing in adults of all ages, linked to rising obesity levels. Evidence shows that the poorer the maternal glucose control in early pregnancy, the higher the baby's risk of developing a congenital anomaly. The National Pregnancy in Diabetes Audit 2016 showed that glucose control was optimal in only about 1/5 of women in early pregnancy with diabetes in Wales.

Pregnancies in all women with pre-gestational diabetes occurring between 1998 and 2015 were identified using hospital data. Using the EUROCAT definition of congenital anomalies the relative risk of having a pregnancy affected by congenital anomaly was calculated and the results are displayed. This shows that Type 1 diabetic mothers have three times the risk and Type 2 diabetic mothers twice the risk of having a baby with a congenital anomaly. The data in Wales confirms the increase in nervous system (including neural tube defects), cardiac, and ear, face and neck anomalies reported in the literature.

Monitoring and appropriate interventions such as increased folic acid pre-conception and during early pregnancy can reduce the risk of a congenital anomaly in this group of women.

Congenital Cardiac Anomalies

The system most commonly affected by congenital anomalies is the cardiac system, and this is the focus of this year’s report and annual meeting. The following are six serious cardiac anomalies with detailed information in the table.

Coarctation of the aorta: an obstruction in the descending aorta.

Transposition of the great vessels: the aorta exits from the right ventricle and the pulmonary artery from the left ventricle.

Fallot’s tetralogy: ventricular septal defect, pulmonary stenosis, right ventricular hypertrophy and overriding aorta.

Hypoplastic left heart syndrome: hypoplastic left ventricle with aortic and/or mitral valve atresia.

Double outlet right ventricle: both great arteries connect (in whole or in part) to the right ventricle often with a rudimentary left ventricle.

Truncus arteriosus: originates from both ventricles of the heart to supply directly the aortic, pulmonary and coronary arterial circulations.

Detection rates for serious cardiac anomalies continue to improve and Wales has some of the best results in the UK. We monitor these rates working closely with Antenatal Screening Wales.

Data from CARIS contributes to international research on medications taken in pregnancy (EUROmediCAT) that can cause fetal cardiac problems e.g. certain seizure medications and some acne treatments. The long-term outlook for children achieving their first birthday is good, although they will need cardiology follow up throughout their lives.

Key data for Cardiac Anomalies, numbers, percentage and detection rates, Wales, 1998-2016

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>All cases</th>
<th>Liveborn cases</th>
<th>Surviving to 1 year of age</th>
<th>Anomaly scan (18-20 week) detection rate 2014-16</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coarctation</td>
<td>360</td>
<td>324</td>
<td>93.4</td>
<td>90.4</td>
</tr>
<tr>
<td>Transposition</td>
<td>237</td>
<td>176</td>
<td>77.9</td>
<td>88.6</td>
</tr>
<tr>
<td>Fallot’s</td>
<td>232</td>
<td>207</td>
<td>95.0</td>
<td>93.7</td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>195</td>
<td>86</td>
<td>45.5</td>
<td>58.1</td>
</tr>
<tr>
<td>Double outlet right ventricle</td>
<td>134</td>
<td>97</td>
<td>76.4</td>
<td>80.4</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>66</td>
<td>42</td>
<td>65.6</td>
<td>73.8</td>
</tr>
</tbody>
</table>

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* Pregnancies ending in 2016 were not included in the survival analysis as it is not yet possible to determine whether a live birth survived to one year of age.

* Based on five years (2012-2016) due to small numbers.