

**Results** 130 cases met the case definition with an overall annual incidence of 5.04 cases per million children under 16 years.

London, East Midlands, West Midlands and Scotland had estimated incidences above the national incidence. Boys (91/130; 70%) were significantly more affected than girls (39/130; 30%) and the majority were of Black (44.6%) and South Asian (36.2%) ethnicity with a median age of 18 months. The commonest clinical presentations were bowed legs, swollen wrists and radiological rickets. Comorbidities included fractures (15/130; 11.5%) hypocalcaemic seizures (11/130; 8.5%), and dilated cardiomyopathy (4/130; 3%) Two children died of dilated cardiomyopathy from vitamin D deficiency. The commonest associated conditions were cows milk protein allergy (19/51; 19%; ) iron deficiency (8/51; 7%) and eczema (8/51; 7%) At the time of diagnosis 77% of children were not receiving vitamin D supplements. 19 children had rickets despite being reported to be receiving appropriate supplementation. All confirmed radiological cases had either high parathyroid hormone and/or low phosphate. Following diagnosis, most clinicians initially prescribed treatment themselves, with huge variation in duration of prescriptions. In a further 10 cases, rickets was confirmed but excluded in the incidence analysis, for not meeting the case definition (specifically Vitamin D < 25 OHnmol/L), suggesting both dietary calcium deficiency and vitamin D insufficiency as role-players in the presentation of NR in the UK.

**Conclusions** NR continues to affect children in the UK with serious sequelae. Uptake of vitamin D supplementation remains low and constitutes a failure of current public health policy. A UK national policy focusing on vitamin D and calcium supplementation and adherence is required to eliminate this entirely preventable condition.

### 117 REDUCED RATES OF JUVENILE ONSET RECURRENT RESPIRATORY PAPILLOMATOSIS IN AUSTRALIA AFTER IMPLEMENTATION OF A NATIONAL HPV VACCINATION PROGRAM

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**Aims** Juvenile onset Recurrent Respiratory Papillomatosis (JoRRP) is a rare chronic disease caused by human papillomavirus (HPV) types 6 and 11. Children with RRP require multiple surgical interventions; tracheostomy may be needed and sometimes the disease is fatal. Infections are now preventable through HPV vaccination. Following an extensive quadrivalent HPV vaccine catch-up program for females aged 12–26 years

in 2007–2009, in Australia, we aimed to monitor the changes in incidence and demographics of JoRRP over time.

**Methods** The Australian Paediatric Surveillance Unit (APSU) conducted national surveillance for JRRoP using its well established reporting system. In addition to the ~1450 paediatricians who report to the APSU each month, paediatric otorhinolaryngologists were also enrolled in the APSU and offered HPV typing. We report findings for the five-year period to end 2016.

**Results** The average annual incidence rate was 0.0715 per 1 00 000 children aged <16 years. The largest number of cases was reported in the first year, with decreasing annual frequency thereafter. The rate declined significantly from 0.163 per 1 00 000 in 2012 to 0.024 per 1 00 000 in 2016 ( $p=0.034$ ). Among the 15 incident cases 60% male, 60% were first born children and 13 (87%) were born vaginally. None of the mothers of these children had received the HPV vaccine before pregnancy, and 3 (20%) of the mothers had a history of genital warts. Seven genotyped cases were positive for HPV including 4 that were HPV6 positive and 3 that were HPV11 positive.

**Conclusion** To our knowledge this is the first report internationally documenting decline in JoRRP incidence in children following a quadrivalent HPV vaccination program.

### 118 THE NATIONAL CONGENITAL ANOMALY AND RARE DISEASE REGISTRATION SERVICE (NCARDRS): THE FIRST YEAR

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**Background** Congenital anomalies (CA) cause around a fifth of infant deaths and are a major contributor to subsequent illness and disability. Regional registers of CA have existed for over 30 years but an effective national registration system has long been needed. More recently, in recognition that collectively Rare Diseases are thought to affect up to 3.5 million people in UK, rare disease registration has been planned which will align with parallel European initiatives.

**Methods** From April 2015 Public Health England launched NCARDRS, incorporating the 7 existing regional CA registers and the National Down Syndrome Cytogenetic Register. Those regions not previously covered (51% of England) were added. A central database was developed for the accrual of new cases and into which to import data held in the pre-existing registers. New data sources for case ascertainment and import of supporting data, such as cytogenetic laboratory feeds, links to BadgerNet neonatal, and Hospital Episode data, have been developed. Pilot work and database development for Rare Disease ascertainment is ongoing. The analysis we report was based on data aggregated from 4 of the pre-existing NCARDRS regions.

**Results** There were 2905 cases with at least one congenital anomaly among 1 41 474 (21%) of births in England in 2015. CA accounted for 17% of infant deaths, half of which were from cardiac anomalies. Two thirds of anomalies were diagnosed prenatally; of those diagnosed postnatally for which there was information on timing, three quarters were diagnosed in the first postnatal week. Rates of non-genetic