

**Conclusion** Chest X-ray is a readily available and commonly performed initial radiological investigation in newborns. Although it can detect most common conditions presenting in newborns, it is important to recognize its limitations and consider further imaging as highlighted in our case series.

# 616 MORBIDITY DUE TO ACUTE LOWER RESPIRATORY INFECTIONS (ALRI) IN CHILDREN WITH BIRTH DEFECTS: A TOTAL POPULATION LINKED DATA STUDY

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**Background** Acute lower respiratory infections (ALRIs) remain the leading cause of hospitalisation among children < 2 years old. Birth defects occur in 5% of live births in Western Australia (WA), and are a major cause of childhood morbidity and mortality. However ALRI hospitalisation rates among children with various birth defects are unknown.

**Methods** We conducted a retrospective cohort study of 245,249 singleton births in WA (1996–2005). Population-based hospital morbidity data and the WA Register of Development Anomalies were linked through the Western Australian Data Linkage System to investigate ALRI hospitalisations in children with and without birth defects. We used negative binomial regression to estimate incidence rate ratios (IRR) for the association between birth defects and number of ALRI hospitalisations over the first 2 years of life, adjusting for known risk factors.

**Results** Overall 11% of non-Aboriginal children and 40% of Aboriginal children with birth defects had a least one ALRI admission before age 2 years. In adjusted analyses, Aboriginal and non-Aboriginal children were more likely to be hospitalised for ALRI if they had a birth defect than children with no birth defects (IRR 2.29, 95% CI: 1.89, 2.78; IRR 2.00, 95% CI: 1.84, 2.17 respectively).

**Conclusions** WA children < 2 years with birth defects are at greater risk of morbidity due to ALRIs, when compared to children with no birth defects. Risk of ALRI hospital admission varies between different birth defect categories.

# 617 FLEXIBLE BRONCHOSCOPY IN THE ACUTE MANAGEMENT OF CONGENITAL LOBAR EMPHYSEMA

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**Introduction** Although uncommon, congenital lobar emphysema (CLE) is a potentially life threatening pulmonary abnormality affecting infants. Lobectomy, sometimes done under emergency conditions, is the universally accepted treatment of CLE with severe symptoms. However, in the developing world trained pediatric thoracic surgeons are not everywhere available. On the other hand, the use of flexible fiberoptic bronchoscopy (FFB) in children has shown an excellent safety record. Furthermore, it can be performed under sedation in most cases. We present here a case of CLE, in which we had been able to relieve the acute respiratory distress using FB.

**Case presentation and procedure** A distressed 4.5 months female infant was referred to us for repetitive prolonged bronchopneumopathy since birth. Successive chest X-rays showed increasing expansion and hyperlucency of right hemithorax, mediastinal shift, and compression of the left lung. At FFB the apicoposterior segmental bronchus of the right upper lobe (APSB/RUL) was narrow, flaccid and showed an expiratory check valve obstruction. By manipulating and rotating the bronchoscope tip into APSB/RUL and applying suction, we had succeeded to release the trapped air in

the RUL. Clinical and radiological manifestations resolved completely following the procedure.

**Conclusions** In addition to the role it might play in the diagnostic workup of patients with CLE, FFB can be used as a tool for relief of obstruction. In certain situations, this may be life saving, especially in places and settings where emergency lobectomy cannot be arranged.

# 618 BENEFIT OF PRENATAL DIAGNOSIS OF COMPLEX CONGENITAL HEART DEFECTS ON NEONATAL MORBIDITY

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**Background and Aims** As survival rates in neonatal cardiac surgery improve over time, morbidity becomes more important in complex congenital heart disease (cCHD). We investigated whether prenatal diagnosis of cCHD affects preoperative and early postoperative morbidity in neonates.

**Methods** Retrospective observational study, performed between July 1<sup>st</sup> 2004 and July 1<sup>st</sup> 2011, including 247 infants with cCHD requiring surgery within the neonatal period. 113/247 patients were diagnosed with cCHD by prenatal screening (PreDx). Morbidity was based on preoperative use of high dose prostaglandin E and pre- and postoperative cardiac function, ventilation time, length of admission (LOF), inotropic support requirement and presence of severe complications (resuscitation, acute kidney and liver injury, and neurological events).

**Results** Complexity of cCHD was higher in children diagnosed PreDx. After correction of complexity, median pre-operative ventilation time (3 vs 0 days), LOF (7 vs 9 days) and preoperative cardiac function were significantly better in PreDx neonates; and correction of acidosis, preoperative inotropic support and high dose prostaglandin E was significantly less common. Severe complications occurred significantly less when PreDx. There were no significant differences in postoperative morbidity and overall mortality.

**Conclusions** Prenatal diagnosis of cCHD improves pre-operative morbidity significantly. The patient condition is better compared to postDx and the need for preoperative intensive care treatment is less. Seemingly, intensive care treatment can achieve a good pre-operative condition, even in neonates deteriorated due to an at birth unknown cCHD. This could explain why post-operative morbidity was equal in both groups.

# 619 AN OUTBREAK OF NEURAL TUBE DEFECTS IN IRAQ: A CASE REPORT

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**Aim** The aim of this study was to report an outbreak of Neural Tube Defects (NTDs) in Iraq, and to highlight the possible responsibilities of international and local authorities for an action for the control of this outbreak in this region.

**Methods** Information for the occurrence of NTDs was gathered from reports published from an Iraqi western region, and for other countries from reports published by the International Clearinghouse for Birth Defects and European Network for Surveillance of Congenital Anomalies.

**Results** Prevalence rate of NTDs was 33 (per 10,000 births, CI95%: 21–44) in Iraq while different rates have been reported from various parts of the world ranging from 12.6 (per 10,000 births) in Cuba, 9.6 (per 10,000 births) in Norway, 8.7 (per 10,000 births) in China, 7.03

(per 10,000 births) in the neighboring country of Iran and 4.9 (per 10,000 births) in Hungary. These figures show that an "outbreak of NTDs" is seemingly being occurred in the Iraqi region of Al-Ramadi in the west of the country. The rate of NTDs in this region of Iraq is about 2.6, 3.4, 3.8, 4.7 and 6.7 times higher than that of reported from Cuba, Norway, China, Iran and Hungary, respectively. It is also 3.2 times higher than that of estimated/expected for the global population.

**Comments** The very high occurrence of NTDs in this Iraqi region indicates that there is an urgent need for an action by regional health authorities and international agencies to the control of this outbreak in the area.

## 620 THE PATTERN OF NEURAL TUBE DEFECTS IN A HIGHLY ENDOGAMOUS SOCIETY: 25 YEAR INCIDENCE TRENDS

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**Objective** The aim of this study is to determine the incidence and trends of NTD over a period of 25 years in the State of Qatar.

**Design** This is a retrospective hospital based cohort study.

**Setting** The survey was carried out in the main territory hospital in the State of Qatar.

**Subjects and methods** The study was conducted from 1985 to 2009 with a total of 302,049 newborns at the Women's Hospital in Qatar screened for NTD. NTD were defined according to the International Classification of Diseases, Tenth Revision (ICD-10). Study parameters included age, gender, ethnicity, parental consanguinity, and residential area.

**Results** The combined prevalence of NTD (total myelomeningocele and anencephaly) during the 25 years period was 1.09 per 1000 births. The prevalence of anencephaly was 0.36 per 1000 births. There were 131 (42.1%) males and 180 (57.9%) female newborns with NTD, corresponding to incidence of 0.899 and 1.289 per 1000 in male and female newborns, respectively. This difference was significant ( $p < 0.001$ ). Consanguinity was seen in 36.7% of the parents.

**Conclusion** The present study revealed that Qatar has a relatively low incidence of NTD which is comparable to neighboring countries in the Arabian Gulf region. High rates of consanguinity and lack of periconceptual folic acid intake among mothers appear to be the major factors contributing to NTD in Qatar.

## 621 POLYMORPHIC LENGTH OF *FOXE1* ALANINE STRETCH IN ISOLATED CLEFT LIP AND/OR PALATE

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**Background and Aim** Isolated cleft lip with/or cleft palate (CL/P) is a common complex birth defect that varies in prevalence with Asian and Amerindian ancestry having the highest rates. While several genes had significant association to the CL/P, the *FOXE1* gene is involved in embryonic formation and is expressed in the secondary palate epithelium in human fetus. Since the first description of *FOXE1* mutation found in cases of cleft palate, thyroid agenesis and choanal atresia, Moreno had conducted a genome-wide scan for CL/P patients and confirmed the highly linkage to the 9q23–33, which resided the potential *FOXE1*. Herein, we are trying to investigate the isolated CL/P in Taiwan to see whether the polymorphic length of *FOXE1* play an important role in the palatogenesis.

**Method** Eighty patients with isolated CL/PCL/P and one hundred controls were recruited in the study. Genomic DNA was amplified by PCR the amplicons containing polyalanine tract (234 to 258 bp; 11–19 alanines) were purified, then directly sequenced.

**Results and conclusion** The 14/14 genotype in polymorphic alanine stretch was most frequent both in cases (98.7%) and in controls (98.0%), whereas the heterozygous 14/16 accounted for one case and two controls. The 14 alanine stretch accounts for the major allele frequency of the polymorphic length in *FOXE1* which consists of the high frequency in previous report from Japan. Although CL/P patients was linked to *FOXE1*, the polymorphic *FOXE1* alanine stretch has no association with isolated CL/P in Taiwan. It appears to reflect the heterogeneity in formation of CL/P, perhaps a population difference.

## 622 CONGENITAL CYSTIC ADENOMATOID MALFORMATION: IS IT ALWAYS BAD NEWS?

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**Aim** To evaluate whether conservative approach to management of asymptomatic neonates with antenatal diagnosis of congenital cystic adenomatoid malformation of the lungs (CCAM) is safe and appropriate.

**Method** We undertook a retrospective review of all cases with antenatal diagnosis of CCAM from 2004–2010 in a lead perinatal centre. Pertinent data was extracted and the outcome and management of all affected infants were reviewed up to 1 year of age.

**Results** Twenty-six pregnancies were complicated with CCAM. Three pregnancies were excluded: two terminated and one fetus had bronchogenic cyst. Twenty-three singleton pregnancies were included in the study. There was a threefold increase in CCAM in the last two years of the study as compared to the first two years. CCAM lesions underwent complete resolution in four (17%) fetuses, partial resolution in 12 (52%), remained static in four (17%) and increased in one (4%) fetus. Eleven (48%) fetuses did not have any associated complications. 15 (65%) infants were asymptomatic at birth and eight (35%) were symptomatic. In two symptomatic infants respiratory distress resolved spontaneously. Three symptomatic infants had surgery and one died. A further three symptomatic preterm infants died due to extreme prematurity. All the surviving, non-operated infants: 17 (74%) were followed up and none required hospitalization or surgical interventions in infancy.

**Conclusions** Our study shows that conservative management of asymptomatic infants with CCAM; consisting of symptoms surveillance, radiological investigations and consideration for surgery if symptoms arise and persist is safe and may be more appropriate to elective surgery in infancy.

## 623 GASTROSCHISIS TRANSFERS CONDUCTED BY THE WEST MIDLANDS NEONATAL TRANSFER SERVICE

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**Background and Aims** Gastroschisis has an incidence of approx. 3 per 10,000<sup>1</sup> with an increasing trend in the UK.<sup>2</sup> Gastroschisis requires immediate postnatal surgical care. Within the West Midlands, newborns with gastroschisis are resuscitated and transferred by WMNTS to a surgical centre. Network guidelines suggest completion of transfers within 4 hours of birth.

**Methods** Retrospective review of gastroschisis transfers conducted by the WMNTS from Jan 2008 to Dec 2011.