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Background Fetal Alcohol Syndrome (FAS) comprises a triad of growth impairment, central nervous system dysfunction and characteristic facial features. Diagnosis is complex and often not recognised at an early age. The three facial features: short palpebral fissures, smooth philtrum and thin upper lip, are unique to FAS. Clinical examination is inherently subjective and apart from palpebral fissure length, minimal reference data is available in neonates. Establishing a standardised method and normal range would promote an objective assessment. Earlier diagnosis would enable earlier effective interventions.
Methods Standardised digital facial photographs were taken of normal term Caucasian neonates. Mothers completed anonymous questionnaires about alcohol consumption during pregnancy. Photographs were assessed using Facial Analysis Software to obtain values for palpebral fissure length (PFL) and upper lip circularity (LC). Upper lip thinness and philtrum smoothness were ranked according to 5-point Likert Scale.
Results 29 infants were studied, 17 male: 12 female. Mean gestational age 40.3 weeks (range 37.1–42.3), mean weight 3556g. 23 (79%) had no prenatal alcohol exposure whilst 6 had minimal exposure (1–2units/week). PFL measurements could be obtained from 21 photographs (72%) with mean of 15.6 mm (range 13.7–18.7mm). Upper lip and philtrum values could be determined in 24 (83%). Mean LC was 57.21 (range 31.4–128.2). Mean rank scores for upper lip and philtrum were both 3.
Conclusion It has been possible to gain measurements of facial features in just over ¾ of neonates studied, showing the feasibility of this technique in this age-group. Further results are needed to establish reference ranges.

613 CONGENITAL DIAPHRAGMATIC HERNIA SURVEILLANCE IN IRELAND

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Introduction The incidence of Congenital Diaphragmatic Hernia (CDH) varies from 1 in 2,000 to 15,000 births per annum. Due to the legislation prohibiting termination of any pregnancy in the Republic of Ireland (ROI) we hypothesised that the incidence would be different to international reports. There is no mandatory reporting of CDH cases in the ROI and Northern Ireland (NI).
Aims To determine the true incidence of CDH in the ROI and NI.
Methods Reporting of CDH cases to the Irish Paediatric Surveillance Unit (IPSU) based in Dublin started in January 2010. Doctors reported new cases to IPSU using the pro-forma provided. Details of the reported cases were collected using a detailed CDH questionnaire.
Results 23 cases were reported to the IPSU within the 2 year period consisting of 10 cases in 2010 and 13 in 2011; 8 cases from NI and 15 cases from ROI. The incidence rate in the 2 years using IPSU figures was 0.12 per 1000 live births per annum. The questionnaire was completed in 12 cases. 11/12(92%) made it to the surgical centres. However the Hospital in Patient System record in one of the three surgical centres indicated that only 6/15(40%) cases managed in that centre in the 2-year period were reported to IPSU.
Conclusion It was difficult to determine the true incidence of CDH using the IPSU data due under-reporting. Increased reporting may be achieved by raising awareness and by the use of capture-recapture mechanism. The establishment of a national congenital anomaly register is also indicated.

614 THREE SIBLINGS WITH NEONATAL PRESENTATION OF GRISCELLI SYNDROME

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Introduction Griscelli syndrome type 2 is a rare disorder charecterised by pigment dilution (silvery hair), variable immune deficiency, and tendency to develop a life threatening hemophagocytic syndrome. Presentation in neonatal life is even more rare.
Materials and Methods These are three siblings to first cousin parents who presented with Griscelli syndrome in the neonatal life with dessimel outcome.
Results J, A, S, are three siblings, two sisters and one brother born to first cousin parents. All had normal vaginal full term delivery. All presented on day one of life with pallor, silvery hair, and hepatosplenomegaly. Investigations revealed variable degrees of anemia & thrombocytopenia. BM revealed hemophagocytic syndrome. There was no HLA matched BM donor so they were treated conservatively. They had repeated admissions for infections and received several blood and platalet transfusion. They had stoem courses and died by two months of age.
Conclusion This is probablly the largest series of Griscelli syndrome presenting in neonatal life. BM is the only hope fore this syndrome.
Athorough family history is always helpfull in diagnosing difficult cases.

615 CONGENITAL LUNG MALFORMATIONS PRESENTING WITH SIMILAR CHEST X-RAY CHANGES AT BIRTH

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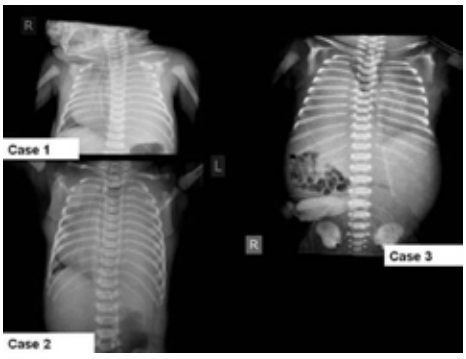
Congenital defects of diaphragm or malformations of lung usually present in first few hours of birth and if left undiagnosed can lead to significant morbidity and mortality. Early accurate recognition is paramount for subsequent management.
Aim To highlight cases presenting to tertiary neonatal unit with respiratory distress on admission and similar chest x-ray changes.
Case 1 Term infant, one of dichorionic diamniotic twins with antenatal history of polyhydramnios admitted with respiratory distress soon after birth. Initial Chest X-ray (CXR) showing homogeneous opacification of left hemithorax with mediastinal shift to right. Subsequent CT chest revealed Bronchogenic cyst.
Case 2 Term infant admitted with respiratory distress. Initial CXR showed homogeneous opacification of left hemithorax with mediastinal shift to right. Subsequent CXR revealed left sided diaphragmatic hernia.
Case 3 Premature infant born at 34 weeks of gestation with multiple congenital anomalies. Initial CXR showed almost complete opacification of right hemithorax. Chest ultrasound was suggestive of severe right sided diaphragmatic eventration.



Abstract 615 Figure 1 Initial Chest X-ray