Pre-piloted forms were used by one reviewer and cross verified by second reviewer. Included studies were assigned a grade based upon their level of evidence and critically appraised using defined tools. Data were not synthesized for metaanalysis due to nature of literature reviewed and heterogeneity.

**Results** 3,592 titles were screened with 43 full-texts assessed for eligibility. 30 studies were deemed eligible for inclusion. These included 19 cohort studies, two qualitative studies, seven cross-sectional studies, one systematic review (peer reviewed articles up to January 2011) and one randomised control trial. Grey literature search from guidelines, catalogues and repositories yielded additional 10 guidelines.

**Conclusions** Optimally designed neonatal unit has many possible health implications, including improved breastfeeding, infection and noise control, reduced length of stay, hospitalisation rates and potentially improved neonatal morbidity and mortality. ‘Single family room’ design for neonatal units is recommended. Consideration should be given to infection prevention and control, including sink frequency and positioning, and airborne isolation facilities. Support areas for families, staff and breastfeeding mothers are also recommended. High quality, family centred neonatal care could be assisted through well grounded, future proofed and technology enabled design concepts that have the potential to impact upon early life development. Nature of the topic poses inherent limitations for conduct of randomized trials; however observational studies using standardised methodologies could add further evidence.

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**P481 CLINICAL CASE OF THE NEW MUTATION IN PIGW GENE WITHOUT HYPERPHOSPHATASEMIA**

1Dinara Sadykova, 1,2Eugenia Slastnikova*, 1Svetlana Zdanova, 2Zulfiya Vafina, 2Zulfiya Khabibrakhmanova. 1Faisel He Kazan Smu Moh, Kazan, Russian Federation; 2Children’s Republican Clinical Hospital Of The Republic Of Tatarstan Ministry Of Health, Kazan, Russian Federation

The 6 day old girl was born at the gestational age of 37 weeks, from the 6th pregnancy, 3rd urgent delivery; her mother’s age was 37 years. The first child was born in 2002 and died at the age of 3 months, the second child was born in 2004, died at the age of 6 months, the 3rd pregnancy in 2008 resulted in antenatal fetal death at the gestational age of 22 weeks, the 4th and the 5th pregnancies were medically interrupted at early stages on request of the family. Convulsive syndrome was observed in 2 eldest children, usage of the valproic acid drugs as an anticonvulsant therapy was ineffective. The patient has convulsive syndrome, with an increase in dynamics. Eye recti; genital hypoplasia. From the first days of life the child is deformed; hands - flexion contracture of the fingers, feet - equinovarus. Phenotypically, the girl is similar to the previous children:

- macrosomia; low foreheaded skull, supraorbital arc; face - hypertelorism, saddle nose, macrognathia, macroclogalia; ears are deformed; hands - flexion contracture of the fingers, feet - placing the fingers on each other; frog belly, diastasis of recti; genital hypoplasia. From the first days of life the child has convulsive syndrome, with an increase in dynamics. Eye phenomena prevailed, head dropping backwards, anxiety expressed. On 8 day of life phenobarbital was prescribed. While receiving phenobarbital, the intensity of convulsions somewhat decreased, however, it was not possible to achieve complete relief from both the clinical picture and the EEG data. The treatment included the drug Levetiracetam (Keppra), the dose increased to 40 mg/kg/day. Against this background, convulsive episodes remain. Homozygous c.1463G>T p. Cys488Phe mutation was detected in the PIGW gene (NM_178517.3) on chromosome 17q12, which encodes the synthesis of glycosylphosphatidyl inositol. This mutation is absent in population databases (EXAC, GnomAD, GenOMED), but several computer algorithms predict its potential pathogenicity. In our patient, alkaline phosphatase activity remained within the normal range. It’s described in literature that clinical manifestations similar to our patient. Thus, one of the possible causes of intractable convulsive syndrome, accompanied by characteristic phenotypic signs and, not always, high alkaline phosphatase activity, should be sought in the group of diseases caused by the biosynthesis glycosylphosphatidylinositol disease. Hopefully, over time, specific therapy will appear for these patients.
intrauterine fibroids, myomas or the foetal limbs could result in deformation.

In the case described the history of oligohydramnios may have contributed to skull depression by bringing the foetal skull into contact with the solid structures within the maternal pelvis.

Proposed treatments for congenital SD include conservative management, surgical and non-surgical interventions. The majority of skull depressions resolve spontaneously, therefore as in this case, in the absence of neurological symptoms a conservative approach of a six-month observation period is recommended.

P483 HOME OXYGEN REFERRAL IN PREMATURE NEONATES BORN IN UMHL OVER TWO YEARS PERIOD

Aisha Ijaz*, Mohammed Abu Bakar, Rizwan Khan, Ghulam Raza. University Maternity Hospital, Limerick, Ireland

Background: Bronchopulmonary dysplasia (BPD) has been a challenging condition for neonatologists (1). Prematurely born infants who had BPD may require supplementary oxygen at home for many months (2).

Objectives: To determine the number of premature babies discharged on home oxygen over two years period in UMHL and to confirm about average gestation age at which babies were self ventilating in air.

Methodology: Retrospective data of 38 babies equal to or less than 29 weeks of gestation was collected, born in UMHL from February 2016 to February 2018. Different variables were studied including Gestation at birth, Birth weight, Date of admission, Date of discharge, Gestation for self ventilation in air, Gestation of home oxygen referral and gestation at discharge. Management of Chronic Lung Disease in the form of steroids, diuretics, patent ductus arteriosus (PDA) treatment and number of blood transfusions were also looked upon.

Results: Total 38 babies born in UMHL from February 2016 to February 2018, equal to or less than 29 weeks of gestation. 26%(n=10) babies were excluded from the study including 21%(n=8) babies died, 3%(n=1) chart missing and 3%(n=1) baby lost follow up. 71%(n=20) were born in the gestation range of 26 to 28 weeks and 70%(n=19) were between birth weight of 750 grams to 1250 grams. 59%(n=16) babies were self ventilating in air between 31 to 35 weeks of gestation compared to 11%(n=3) who took over 41 weeks of corrected gestational age. 4%(n=1) babies were not able to wean off from oxygen over two years period. 29%(n=8) of the babies didn’t need any intervention (Diuretics, Steroids, PDA management, Blood transfusion) during their stay in NICU while 29%(n=8) needed just one intervention. 43%(n=12) of the babies needed two or more interventions to achieve self ventilation in air.

Conclusion: Only one baby out of total 28 was discharged on home oxygen over two years period and most of the babies were able to achieve self ventilation in air between 31st to 35th weeks of gestation.

REFERENCES