Maternal phenylketonuria, newborn screening program, developmental delay.

P434 TYROSINEMIA TYPE 1: A CASE REPORT

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10.1136/archdischild-2019-epa.770

Introduction Tyrosinemia type 1 is an AR inherited metabolic disorder attributed to deficiency of Fumarylacetoacetatehydrolase, which is a terminal enzyme in the metabolism of tyrosine. The gene for this enzyme has been mapped to the long arm of chromosome 15. Its prevalence has been reported as 1: 100.000. The patients with tyrosinemia expire in the early years of their lives. There is a markedly increased risk of hepatocellular carcinoma among the survivors.

Case report An Saudi 8-month-old male infant presented with complaints of abdominal distension, fever, jaundice, melena and disturbed level of consciousness for 3 days prior to admission to PICU (in King Fahd Hospital, Al-Baha). He was the 1st child of 1st degree consanguineous parents.

His motor and mental development were delayed. One day after admission, the infant developed repeated daily attacks of neurological crises in bouts of irritability, crying with increased tone and deep tendon reflexes in the lower limbs.

On physical examination patient looked sick, pale, jaundiced and drowsy. He had rachitic signs. There was mild lower limb edema. Fine crackles were audible bilaterally on the chest. Abdomen was distended. Liver was palpable 5 cm below right costal margin. Spleen was just palpable below left costal margin. There was positive shifting dullness, with scrotal edema. The infant was drowsy, with poor vision, and hypertonic, with exaggerated deep tendon reflexes.

Hematological screening CBC showed anemia and thrombocytopenia. CRP was positive and urine analysis showed albumin & RBCs.

Biochemical examination showed that BUN: 12.5 mmol/L, creatinine 27 µmol/L, glucose: 2.9 mmol/L, Ca: 2.2 mmol/L, phosphorus: 0.64 mmol/L, Mg: 0.57 mmol/L, albumin: 28 µmol/L, T. bilirubin: 35 µmol/L, direct bilirubin: 17 µmol/L, alkaline phosphatase 426U/L, AST: 65U/L, gamma GT: 130U/L, LDH: 566U/L, ammonia: 121 µmmol/L. Coagulation screening was abnormal. Blood Alpha fetoprotein was very high, ABG examination showed compensated metabolic acidosis.

Tracheal aspirate C/S showed MRSA The left wrist graphy showed evidence of rickets. Abdomen ultrasonography revealed hepatosplenomegaly with moderate ascites. Liver examination showed multiple hyperechoic masses. Abdominal CT showed hepatomegaly with multiple hyperdense masses, splenomegaly and ascites. CT brain showed cerebral atrophy.

Blood phenylalanine, tyrosine and methionine levels were all high. Urine examination showed increased levels of organic acids.

So, the infant was diagnosed as tyrosinemia type I and was prescribed a phenylalanine and tyrosine restricted diet (special formula milk) and received treatment for Vit. D resistant rickets. Following treatment, patient showed much clinical and laboratory improvement, but not to the safe level to conduct liver biopsy.

P435 RETINOPATHY OF PREMATURITY PREVALENCE AND RISK FACTORS IN DEVELOPING COUNTRIES

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10.1136/archdischild-2019-epa.771

Retinopathy of prematurity prevalence and risk factors in developing countries

Introduction Retinopathy of prematurity (ROP) is an avoidable disorder in preterm infants, which can lead to blindness unless it is diagnosed and treated at early stages.

Objectives The goal of this study is to determine the prevalence of ROP and its predisposing factors in preterm infants admitted to NICU at community hospital, Mashhad, Iran.

Method ROP screening was done for infants that admitted to NICU weighing less than 1500 mg and a gestational age of 32 weeks or less was conducted from January 2018 to 2019 in. A total of 149 neonates (89 males and 60 females) underwent retinal evaluation using indirect ophthalmoscopy during the fourth week and were periodically followed up.

Prenatal risk factors of ROP were evaluated using univariate analysis.

Results Of the 149 neonates under study, 42 were ROP positive out of whom 17 (40%) were at stage 2, 12 (29%) at stage 1, 2 (5%) at stage 3 and 2 (5%) at stage 4 while 19 (21%) of newborns were treated with Avastin injection. The results of univariate analysis showed that there was a significant difference between ROP occurrence and gestational age, birth weight and oxygen therapy duration. ($p = 0.0001$). However, there was no significant relationship between the ROP occurrence and gender ($p = 0.55$), stage of disease and gestation age ($p = 0.208$) and oxygen therapy duration ($p = 0.314$).

Conclusion The estimated prevalence of ROP in this study was 28% that was higher than developed countries. Gestational age, oxygen therapy and low birth weight are known risk factors of ROP. The risk of ROP in a gestational age of less than 32 weeks is 4.2 times higher than the gestational age of more than 32 weeks. Therefore, it is essential to make plan for the most effective control over the predisposing factors like oxygen therapy, preventing premature delivering, and incidence of birth asphyxia in NICU.

Retinopathy, preterm infant, prevalence, developing countries

P436 KNOWLEDGE AND PRACTICE OF PULSE OXIMETRY SCREENING FOR CRITICAL CONGENITAL HEART DEFECT IN NEWBORN INFANTS BY PAEDIATRICIANS IN BENIN

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10.1136/archdischild-2019-epa.772

Introduction Pulse oximetry screening for critical congenital heart defect in newborn infants is not widely used even in developed countries. In Benin, the diagnosis of congenital heart disease is often late, making the surgical treatment impossible

Objective To assess the knowledge and practice of pulse oximetry screening for critical congenital heart defect in newborn infants by paediatricians in Benin.

Method This was a phone survey or a direct interview that interested pediatricians in Benin. The data examined included the seniority of paediatricians, their location, their knowledge and practice of pulse oximetry screening for critical congenital heart defect in newborn infants. The data has been analyzed with EPI info 3.5.4 software.

Results we have identified 84 paediatricians but only 55 (65.47%) participated in this survey. The average duration of paediatric practice was 10.6 ± 8.04 years (1–29). Most of them exercised in LITTORAL district ($n = 34$) and in private clientele ($n = 22$). Respondents reported hearing about pulse oximetry screening for critical congenital heart defect in newborn infants in 65.45% of cases. Only 12.73% were aware of a screening protocol and 16 paediatricians routinely practised it. Only one respondent had a program for screening of critical congenital heart defects by pulse oximetry at its center. The exercise time was inversely associated with knowledge of this screening ($p = 0.005$). This knowledge did not influence the practice of screening critical congenital heart defect in newborn infants ($p = 0.18$).

Conclusion Pulse oximetry screening for critical congenital heart defect in newborn infants is few used by paediatricians in Benin. Its systematization would improve the screening.

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HEALTHCARE PROVIDER'S PRACTICES REGARDING PAIN MANAGEMENT IN NEONATAL DEPARTMENTS

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10.1136/archdischild-2019-epa.773

Pain control is one of the universal strategies for care delivery in all neonatal departments. The problem of pain management in neonates is of great importance for a medical institution, and not only the accountability of individual specialists.

The aim to analyze healthcare provider's awareness about pain management in newborns.

Methods Cross-sectional descriptive study was conducted at in neonatal departments of Perinatal Centre (III level), Irkutsk, Russia, from April to June 2018. Data were collected from 61 respondents: 46 (75.4%) nurses and 15 (24.6%) physicians. The ad hoc questionnaire contained 13 questions regarding respondents' demographics, professional qualification, their knowledge and practices in neonatal pain management. Part of the questionnaire contained information about availability and quality of local pain control guidelines in services. Pain intensity perception during painful procedures was described using a 5-point Likert scale. Data were analyzed using «Statistica 6.0» soft. The level of significance was $p < 0.05$.

Results Most respondents were female ($n = 59$), aged 23 to 60 years, and 56% had 5 to 10-year experience in providing neonatal care.

The majority (88.5%) always evaluated pain in their units using a validated EDIN scale. However 38 (62.2%) respondents doubted the efficacy of this tool.

All professionals considered chest drainage ($p = 1.0$), lumbar puncture ($p = 0.9$), intramuscular injection ($p = 1.0$) to

be the most painful procedures (4–5 – points Likert scale). Statistically significant differences among nurses and physicians were found between their perception of pain intensity during intravenous cannulation and heel pricks ($p = 0.001$; $p = 0.009$, respectively). Physicians considered tracheal suctioning ($p = 0.02$) to be the most painful procedure. All staff considered gastric tube insertion ($p = 1.0$) to be the procedure with minimal pain (1– points Likert scale). All professionals took pharmacological measures for pain relief during chest drainage ($p = 0.8$), tracheal intubation ($p = 0.8$), lumbar puncture ($p = 1.0$). The most reported non-pharmacological measure for neonatal pain therapy was oral glucose both for nurses and physicians (84% vs 75%, respectively). Majority of nurses reported of non-pharmacological pain relief measures used for all routine painful procedures irrespective of pain intensity.

Conclusion The survey showed that the neonatal pain assessment is not systematic, pharmacological measures for pain management are used limitedly due to reservations about analgesic-related neurotoxicity, despite of pain intensity. Non-pharmacological methods are more used according to the staff, especially nurses, reports.

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DESIGN OF AN ASSESSMENT TOOL TO EVALUATE NEONATAL CARE PRACTICE ACROSS A RURAL PROVINCE IN CAMBODIA AND SUBSEQUENT INITIATION OF A COMPREHENSIVE NEONATAL TRAINING PROGRAMME FOR MIDWIVES, NURSES AND DOCTORS

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10.1136/archdischild-2019-epa.774

Introduction The neonatal mortality rate (NMR) in Cambodia is 18 per 1000 births. The NMR in rural areas is higher, with a reported rate of 25 per 1000 in the province being studied. A large-scale programme 'The Saving Babies Lives Programme' is currently being piloted to decrease neonatal deaths in Cambodia. An assessment of the current neonatal care practice was necessary to highlight areas for improvement. Our aim was to create an assessment tool to obtain a comprehensive overview of neonatal services to inform the implementation of a sustainable improvement strategy including a comprehensive training programme for midwives, nurses and doctors.

Methods An assessment tool was adapted from the KAP survey model (Knowledge, Attitudes and Practice). Two further sections were added; Equipment and Staffing, forming a 'KAPES survey'. Tablet-based questionnaires were created using KoboCollect Toolbox. Surveys were written in English and translated into Khmer. National and international guidelines and teaching materials were reviewed to decide core teaching topics and the level of care to be provided at each facility. The research team were trained as instructors and mentors.

Results Knowledge and attitude questions were developed across key domains depending on staff member (midwife, nurse or doctor) in the format of multiple choice questions (MCQs) and statements with a Likert scale. Practice, Equipment and Staffing (PES) included multiple question types and