P31  EVALUATION OF THE RESULTS OF HIP ULTRASOUND SCREENING FOR DEVELOPMENTAL DYSPLASIA OF THE HIP AMONG INFANTS IN A TERTIARY SETTING

1Feza Aydın, 2Emel Gur, 3Tugba Erenar-Ercan*, 4Ahmet Anas. 1Istanbul University, Cerrahpasa Medical Faculty, Istanbul, Turkey; 2Maltepe University, Medical Faculty, Istanbul, Turkey

Background The aim of our study was to investigate the rate of developmental dysplasia of the hip (DDH), and the association between the risk factors for DDH and the results of the hip ultrasound (US) findings among healthy infants

Methods The study group consisted of healthy infants who admitted to the outpatient Healthy Child Clinics of Istanbul University, Cerrahpasa Medical Faculty for their routine control between December 2014 and May 2015. Files of the patients who were followed up at least 1 year of age were reviewed with regard to risk factors (sex, birth weight and length, type of delivery, order of birth, type of presentation, maternal age, history of oligohydramnios, multiple pregnancy, swaddling history, family history of DDH) and hip US findings. All infants had their hip US performed at 4 to 6 weeks of age in the Radiology Department of Istanbul University.

Results A total of 300 infants (175/125: male/female) with a birth weight and length of 3137.03±557.23 gr and 49.69 ±2.68 cm, respectively were enrolled. Fifteen infants had a history of maternal oligohydramnios (5%), 70 (23.3%) were born vaginally, 27 (9%) were born as twins and 2 (0.7%) had a breech presentation. Family history of DDH was present in 17 infants (5.7%) and 28 (9.3%) had a swaddling history. US finding of immature hip was detected in 52.9% of those with a family history of DDH and 13.6% of those with a swaddling history. There was a statistically significant association between family history of DDH and swaddling, and finding of immature hip on US (p=0.0001). In those with findings of immature hip, left hip was affected in 28 (56%) and right hip was affected in 13 (26%) infants. Involvement of left hip was significantly more frequent in case of an immature hip on US (p=0.04). Only one infant had a finding of DDH (Type 2b) on US.

Conclusions The rate of DDH in this study was 0.3%, while the rate of immature hip was 16.7%. Positive family history and swaddling were found to have a strong association with immature hip on US with a significantly more frequent involvement of the left hip. We want to emphasize that family history of DDH should be sought during evaluation of an infant with regard to DDH. We realized that swaddling was still a common practice in Turkey for which the parents should be warned about its associated risk for DDH.

P32  SIGNIFICANCE OF MRI IN THE DIAGNOSIS OF PELVIC OSTEOMYELITIS IN CHILDREN – 2 CASE REPORTS

1Ivan Peychl*, 2Jan Hajicek, 3Jiri Chomiak, 4Jan Lami. 1Ivanka Hospital, Prague, Czech Republic; 2Department of Radiology, Na Bulovce Hospital, Prague, Czech Republic; 3Department of Paediatrics, Na Bulovce Hospital, Prague, Czech Republic; 4Department of Orthopaedic Surgery, Na Bulovce Hospital, Prague, Czech Republic

Background Bone mineral disturbances are often complication of the cystic fibrosis (CF) patients with impact linear growth, quality of life and life expectation and may be contraidication for lung transplant. Osteoporosis is a complex multifactorial diseases which started in the childhood.

The aim of our study was to evaluate bone mineral metabolism in CF children in the Saint-Petersburg.

Materials In the present study 57 CF children, aged 5–18 years were included. For assessment of bone health we evaluated: i) number of significant fractures; ii) dual-energy X-rays and ultrasound examination of the hips were normal. CT scan of the abdomen, pelvis and the left hip showed normal findings on the bones and muscles. We started treatment with bed rest and intravenous ceftriaxon. On day 3, blood culture proved positive for Staphylococcus aureus and the treatment was switched to intravenous clindamycine. MRI of the pelvis and hips performed on day 4 showed changes in the superior ramus of the left pubis and the obturator muscles. Pain in the left hip disappeared within a week of treatment. Laboratory findings showed an increase of CRP level up to 370 mg/l on day 3. The level normalised within 10 days of treatment. The boy was discharged home after 24 days of inpatient stay. The treatment continued with 3 weeks of oral cefuroxim. Follow-up MRI performed 39 days after admission showed partial resolution of the inflammatory oedema of the left superior pubis.

Conclusion Our findings correspond to the 2017 ESPID guidelines: MRI has a primary role in the diagnosis of paediatric acute osteomyelitis. MRI changes of bones and muscles are present in the early stage of the disease and they persist for weeks/months at a minimum. Provided the location of the inflammation is clinically apparent, radionuclide study is not a necessary part of the investigation. CT is of little value in the diagnosis of acute osteomyelitis.
Osteogenesis imperfecta (OI) or Brittle bone disease is a rare genetic connective tissue disorder with the majority of mutations found in collagen type 1 genes (COL1A1/ COL1A2) or their related SIBLING proteins. The condition is characterized by increased bone fragility, resulting from abnormal collagen formation. Recessive forms of OI are associated with increased severity and lethality due to mutations in LEPRE1, encoding prolyl 3-hydroxylase-1 (P3H1) or in CRTAR encoding carilage associated protein. The LEPRE1 gene mutation has been expressed in the Irish Traveller population. We report two children affected with this mutation who have demonstrated a positive response to early bisphosphonate use.

Case 1 A male infant was born in 2013 by elective LSCS at 38 weeks gestation to consanguineous Caucasian parents from the Irish traveller community. Antenatal scans in the third trimester had confirmed multiple in utero fractures. He was commenced on Pamidronate (Bisphosphonate) infusion at four weeks of age. He now sits and transfers independently and is attending school. He has sustained further fractures with trauma.

Case 2 A male infant was born in 2018 by elective LSCS at 39 weeks gestation to consanguineous Caucasian parents from a separate family tribe within the Irish traveller community. Antenatal scans at 22 weeks confirmed limited femur growth and the possibility of skeletal dysplasia were raised at 23 weeks when significant fractures were noted. There was a strong family history of osteogenesis imperfecta with the death of four of his mother’s siblings in childhood with a clinical diagnosis of brittle bone disease. She was believed to be unaffected having never sustained a fracture. He was commenced on an initial dose of zoledronic acid on Day 7 of life with a subsequent reduction in analgesia requirement and increase in peripheral limb activity. He was discharged home at 4 weeks of age.

Discussion Molecular genetics confirmed a diagnosis of type VIII OI caused by a pathogenic mutation in the LEPRE1 (P3H1) gene in both cases. Skeletal survey at birth showed thin ribs, multiple wormian bones and healing fractures of the clavicles, ribs, humeri and femori. Both exhibited a