ray absorptiometry (Lunar iDXA 16[SP2]) with pediatric database in lumbar spine (L1–L4) and in total body less head (TBLH); iii) bone age with Greulich-Pyle method; iv) Tanner's puberty stage and anthropometry. In the cases with significant growth delay or acceleration the height- or biologic age standardization for bone mineral density (BMD) applied

Results The BMD Z score -1SD to - 2SD was in 15 (26.3%) children and Z score < - 2 SD in 5 (8.8%). All children with BMD L1–L4 Z score <-2SD had growth delay so after applying of above mentioned correction methods where were no patients with severe bone mineral density (Zscore < -2 SD) deficiency. Only 3 children had decreased (near -2SD) BMD in total body less head program. No patients with pathological low-energy fractures were observed.

Conclusion We have not found osteoporotic CF patients. Further investigations required for cystic fibrosis patients with borderline decreased bone mineral density (near or less than - 2 SD).

P34 THE LONG-TERM RESULTS OF SINGLE-EVENT MULTILEVEL SURGERY IN TREATMENT OF CHILDREN WITH SPASTIC DIPLEGIA

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Introduction Cerebral palsy is a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. Single-Event Multilevel Surgery – SEMLS has been used by most surgeons for correction limb deformities in the past decades.

The aim Of our study was to evaluate the long-term results of SEMLS, performed in the Mogilev Regional Children's hospital and in the Republican Scientific and Practical Centre for Traumatology and Orthopaedics.

Materials and methods The long-term results of SEMLS in 19 children with spastic diplegia, operated in 2009–2016 were studied. All the patients had spastic deformities of the hips, knees and ankles on the both sides before surgery. All the patients were GMFCS IV. The mean age at time of surgery was 6,7. 138 procedures were performed. All the procedures were made simultaneously on the both sides by two teams of surgeons. Observational Gait Scale (OGS) and Edinburgh Visual Gait Score (EVGS) were used to determine the effectiveness of treatment.

Results The minimal follow-up was 3 years. The mean improvement by OGS was 8,5, the minimal improvement – 3, maximal – 12. The mean improvement by EVGS was 10,1, the minimal improvement – 6, maximal – 16. All the parents noticed the improvements in child's mobility after surgery. One patient has severe valgus foot deformity and calcaneal gait after anterior Achilles tendon transfer as part of SEMLS in 2015. Six patients underwent additional surgery for correction of the secondary bone deformities later.

Conclusions Brain disorders in cerebral palsy are permanent and as S.Terver said – 'The child with cerebral palsy becomes the adult with cerebral palsy'. For the child with cerebral

palsy the main goals of surgical treatment - improving the child's motor activity and elimination of severe limb deformities.

SEMLS changes the whole pathological stereotype of musculoskeletal balance of extremities. It can improve the child's motor activity, eliminate the severe contractures of joints, provide adequate care for him in a relatively short period of time and save these improvements in the long-term follow-up. Care must be taken in choosing the proper method of surgical correction, especially in treatment of equinus.

P35 IMPROVED SURVIVAL IN RARE RECESSIVE OSTEOGENESIS IMPERFECTA THROUGH MULTIDISCIPLINARY TEAM SUPPORT AND EARLY BISPHOSPHONATE USE

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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 CRTAP, encoding cartilage 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

characteristic flat nasal bridge, mid face hypoplasia, with a disproportionately large head to birth weight and long tapered fingers.

The multidisciplinary team aim to maximize the mobility and functional capabilities of these patients to enable them to achieve normal developmental milestones at their own pace, while improving bone strength through a programme of regular bisphosphonate infusions.

P36 CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS, A DIAGNOSIS NOT TO MISS

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Introduction Chronic Recurrent Multifocal Osteomyelitis (CRMO) is classified as an auto inflammatory disorder.¹ It mainly affects children and adolescents. There are several diagnostic approaches described in current literature. Roderick et al formulate the Bristol diagnostic criteria,² which is based on combination of clinical and radiological findings. CRMO is characterized by episodic skeletal pain and swelling, associated with systemic symptoms such as fever and malaise.³ Plain radiograph demonstrates lytic destruction, with sclerosis within the metaphysis of bone. STIR MRI demonstrates oedema in bone marrow, with lytic lesions and periosteal reaction seen occasionally.⁴ The main stream of management is symptomatic relief using mainly non-steroid anti-inflammatory drugs (NSAID). Most cases of CRMO reported are self-limited and resolve without significant complications.⁵

Case description A 5 year old fit and well boy had a febrile illness lasting for a month. He also complained of anorexia, weight loss and abdominal pain. He also had cervical lymphadenopathy. His symptoms were unresponsive to antibiotics. There was no evidence of infections after investigations. Throughout the next year, he had recurrent fever and started to have lower limb tenderness. He was admitted on multiple occasions. He was reviewed by different sub-specialist and investigated thoroughly. He never had raised inflammatory markers or presence of auto-antibodies. On his whole body MRI scan, he was found to have hyper intensity involving the metaphysis of his upper and lower limb bones. Diagnosis of CRMO was then confirmed by a specialist from tertiary hospital on bone biopsy of lytic lesions.

Discussion CRMO is an uncommon diagnosis, cases are reported to have months to years' time interval between presentation to diagnosis (2). Through this case, we aim to raise awareness amongst clinicians to shorten time to make diagnosis and to avoid unnecessary admissions.

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P37 HYPERMOBILE EHLERS-DANLOS SYNDROME EXPLOSION IN THE UK!!

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Background Ehlers-Danlos Syndrome (EDS) is a heterogeneous group of collagen disorders. There is controversy regarding clinical diagnosis of hypermobility EDS. The Beighton score and the recent international classification of EDS are not validated in children. Clinicians get the impression that there is a substantial increase in frequency of referrals with ?EDS especially for musculoskeletal pain.

Methods In this retrospective cohort study we searched local databases of paediatric rheumatology and genetics department for referrals to the paediatric rheumatologist due to concerns regarding having hypermobile EDS. All clinic letters were carefully examined and data collected for patients age, gender, reasons for referral, diagnosis made by rheumatologist and the management plan. We did a descriptive analysis using population proportion confidence interval.

Results In the paediatric rheumatology department we identified 53 referrals (34 female), median age 11 years (range 9 months – 17 years) from June 2017 to June 2018. 59% had anxious parents with one or multiple psychosocial issues as well experiencing musculoskeletal pain. 12 patients had a family history of hypermobile EDS, three patients were diagnosed with hypermobile EDS in a private clinic, for three patients the GP had suggested EDS, one patient had possible skin features of EDS, parents of two patients had concerns following surfing the internet and one patient had a cardiac and vascular problem possibly related to EDS. The patients were diagnosed being a healthy child (49%), or having hypermobility (28%), arthralgia (8%), mechanical pain (7%), chronic pain (6%) and classical EDS (2%). Management included reassurance and discharge (21%), referral to physiotherapy (76%), referral to psychology (13%), referral to other medical teams (21%). Of all patients 11% were followed up after the initial review. The genetic department had total of 245 referrals from January 2017 to June 2018. The referral included 62 patients with ?EDS (19 paediatrics, 43 adults), 14 with ? hypermobility (12 paediatrics, two adults) and 169 hypermobile patients with ?Marfan syndrome (44 paediatrics, 125 adults). Most referrals did not have a significant family history or clinical features.

Conclusion Particular attention should be devoted to education and information in first line health care as well as to more effective management of musculoskeletal pain and parental anxiety.

P38 OUR EXPERIENCE IN RESTORING HIP STABILITY IN CHILDREN WITH CEREBRAL PALSY

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Introduction and Objective Hip instability in children with cerebral palsy (CP) is a serious progressive problem.