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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10.1136/archdischild-2019-epa.391

Introduction Chronic Recurrent Multifocal Osteomyelitis (CRMO) is classified as an auto inflammatory disorder. It mainly affects children and adolescents. There are several diagnostic approaches described in current literature. Roderick et al formulate the Bristol diagnostic criteria, 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. 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. 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.

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 subspecialists 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.

REFERENCE

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

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10.1136/archdischild-2019-epa.392

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.

REFERENCE
1. Aek Sakalouski, Mikhail Herasimenka, Raman Klimau*, Leanid Hlazkin, Mikhail Mikhovich. Republican Scientific and Practical Centre for Traumatology and Orthopaedics, Minsk, Belarus; Brest Regional Children’s Hospital, Brest, Belarus; Mogilev Regional Children’s Hospital, Mogilev, Belarus
10.1136/archdischild-2019-epa.393

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

Aleh Sakalouski, Mikhail Herasimenka, 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.

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Introduction and Objective Hip instability in children with cerebral palsy (CP) is a serious progressive problem.
Materials and Methods 141 surgical procedures were performed on children 3–16 years old in 2006–2016. The purpose of surgery was eliminating the instability of hip (GMFCS 1–3) and relieving pain (GMFCS 4–5).

Triple pelvic osteotomy (TPO) was performed in 5 cases on 5 patients 6–15 years old; TPO combined with varus-detorsion femoral osteotomy (VDFO) - in 8 cases on 6 patients 7–11 years old. TPO, shortening VDFO and open head reduction were performed in 6 cases on 5 patients 7–13 years old who had hip dislocations.

VDFO combined with Salter’s pelvic osteotomy were performed in 4 cases (4–6 years old).

Adductor myotomy was performed in 99 cases (43 patients bilaterally), it was combined with other soft tissue releases in the lower limbs in 32 cases.

Results Hip stabilization was achieved in all cases using TPO and TPO combined with VDFO. Reoperation was necessary in 3 cases after VDFO. The best results after the soft tissue releases were obtained when the Reimer’s migration index was <30%, although in some cases joint stabilization was achieved after the migration index was > 80%. For this group of patients femoral osteotomy was required in 12 cases, femoral and pelvic osteotomies – in 3 cases.

Conclusions Individual approach with a clear understanding of hip surgery and adherence to the principle ‘one-stage and multilevel correction' are necessary in the treatment of children with CP.

When the Reimer’s migration index is <30% the soft tissues release can provide hip stability, in the most severe cases combination of pelvic and femur osteotomies leads to a better result.

This patient was initiated on oral phosphate and one-alpha vitamin D replacement resulting in healing of rickets and improvement of growth. However, his fatigue remains evident and is associated with poor exercise tolerance and persistence of falls. Intervention of physiotherapy [posture and strengthening] and orthopaedics [epiphysiodesis] have improved gait, straightening of legs and growth. However, patients in the UK have demonstrated normalisation of biochemical, improvement of muscle strength and improved long-term mobility with weekly subcutaneous Burosumab injections which is awaiting approval in Ireland.

Discussion/Learning Points Rickets in Caucasian children in Ireland is usually genetic in origin.

Prompt identification of cause allows for early intervention and reduced skeletal deformity. Burosumab [anti-FGF23], the first approved targeted medication for X-linked hypophosphataemic rickets was FDA approved in 2018 and is awaiting approval in Ireland. Patients in clinical trials have demonstrated normalisation of biochemistry, improved muscle strength and long-term mobility with weekly subcutaneous Burosumab injections. In future it is expected targeted therapy in conjunction with continued multi-disciplinary involvement will have a major impact on morbidity in patients living with X-linked Hypophosphataemic Rickets.

PHOSPHATE: THE FORGOTTEN BROTHER – A CASE OF HYPOPHOSPHATAEMIC RICKETS

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Usage of phosphate in children with hypophosphataemic rickets has improved quality of life and long-term outcomes for children. In adults, causes are most often malignancy or primary hyperparathyroidism. In childhood the aetiologies are diverse, may be age specific and many have an underlying genetic basis.

Case Description An 18 month-old boy in good general health, presented with pronounced bowing of his legs since weight-bearing had commenced, associated with reduced growth velocity, flat feet and unsteady gait. Biochemistry was normal apart from low phosphate and raised alkaline phosphatase. Plain radiograph of the lower limbs demonstrated fraying of the distal medial 1/3rd of the metaphyses of the femora and tibiae bilaterally. This characteristic appearance led to suspicion of hypophosphataemic rickets confirmed by elevated FGF-23 levels and genetic confirmation of a de novo mutation in the PHEX gene.

Hypercalcaemia is an infrequent finding in children. In adults, causes are most often malignancy or primary hyperparathyroidism. In childhood the aetiologies are diverse, may be age specific and many have an underlying genetic basis.

Introduction Hypercalcaemia may be found as an incidental finding with no associated clinical features. When clinical features are present there has usually been an insidious onset over a few weeks. Most frequent findings are lethargy, hypotonia, anorexia, weight loss or failure to thrive, polydipsia, polyuria, vomiting, bone pain, constipation and abdominal pain. In severe cases, renal failure, pancreatitis and reduced consciousness may occur. Drug history must include enquiry regarding intake of alternative medicines. It is important to ascertain whether other family members have had hypercalcaemia, renal stones, parathyroidectomy or features of multiple endocrine neoplasia syndromes. Examination should assess the degree of dehydration, look for features of malignancy bone pain or vertebral fractures and the presence of a rash. Evidence of dysmorphism or skeletal disproportion should also be sought.

Case report A 15 months old girl was reviewed in Paediatrics outpatient with mild gross motor delay, fussy eating, constipation, intermittent episodes of screaming and poor sleep pattern. Serum Calcium was noted to be elevated at 2.81, repeat few weeks later remained same. Further workup was done. Bone profile normal, PTH normal, Vitamin D increase risk of inadequacy. Metabolic workup normal. She was commenced on IV fluids and oral Furosemide to bring Calcium down and later discharged home on oral Furosemide and low Calcium diet. Urinary Calcium was noted to be low. Father’s results...