examination is still rarely being documented across Yorkshire, across disciplines, by doctors at all levels, even when there are clear triggers and red flags for the examination.

We propose a change in the admission documentation regionally to include a specific MSK section for acute admissions.

We plan educational events in the region to improve knowledge of MSK examination.

We plan to re-audit in the region, following the introduction of this intervention.

We propose a national audit should be performed. then more educational measures, directed at all levels of doctors should be driven forward, on a national level, to ensure that this picture improves.

REFERENCES

1. pGALS- A screening examination of the musculoskeletal system in school aged children, Foster HE, Jan dial S, 'Hands On', Reports on the Rheumatic Diseases Series 5, Number 15, June 2008.
2. Rheumatology (Oxford). 2004 Aug; 43(8):1045–9. EPub 2004 Jun 8. More 'cries from the joints': assessment of the musculoskeletal system is poorly documented in routine paediatric clerking. Goff I, Bateman B, Myers A, Foster H

G230 THE USE OF COMPOSITE SCORES FOR THE ASSESSMENT OF JUVENILE IDIOPATHIC ARTHRITIS (JIA) IN A ROUTINE OUTPATIENT CLINICAL SETTING

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Several JIA composite scores have been validated for use in clinical research studies, but the practicality of their use in the routine clinical setting is unclear. Our aim was to determine the completion rate of the 6 item composite disease activity score, the American College of Rheumatology core outcome variables (ACR COV), and to calculate the 4 item composite Juvenile Arthritis Disease Activity Score (JADAS) in a tertiary rheumatology unit outpatient clinic setting.

Methods In this single centre clinical service evaluation, a retrospective case note review of 105 consecutive JIA patients attending outpatient follow up clinics in 2011–12 was undertaken. The completion rate of a standardised ACR COV proforma (present in all case notes) was determined, and JADAS was calculated from additional clinical data. Pearson's correlation and logistic regression were used to assess the impact of individual items on changes in JADAS.

Results 105 children with JIA had 193 clinic visits but complete ACR COV data were found in only 68/193 records (35%). Of the 6 items comprising the ACR COV, the ESR accounted for the majority of missing data. Sufficient data was available to calculate JADAS scores in 22 children with two consecutive outpatient visits. The table indicates that changes in JADAS were most dependent on the physician's global assessment, and least dependent on ESR (Pearson correlation). Stepwise regression showed that the physician's global assessment alone predicted 87.5% of JADAS change and the ESR contributed an additional 3.3%.

Abstract G230 Table 1

Pearson correlation		Stepwise regression	
	Change in JADAS		Change in JADAS
Physician global assessment	0.93	Physician global assessment	87.5
Active joints	0.84	ESR	90.8
Patient global assessment	0.73	Patient global assessment	97.1
ESR	0.64	Active joints	98.8

Conclusion In a routine clinical setting, frequent missing data reduced the potential clinical utility of the ACR COV and JADAS composite scores. We speculate that a composite clinical score which does not rely on recording the ESR may improve completion rates without diminishing clinical utility.

G231 MORTALITY PATTERNS IN CHILDREN WITH JUVENILE DERMATOMYOSITIS: TWO DECADES OF EXPERIENCE FROM A SINGLE TERTIARY CARE CENTRE

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Introduction Juvenile dermatomyositis (JDM), is a chronic inflammatory disease characterised by proximal muscle weakness and pathognomonic rash. Although there have been significant advances in management of JDM, the condition still has significant morbidity and mortality associated with it. Complications like breakthrough infections, gastrointestinal tract vasculitis, interstitial pneumonitis and myocarditis pose significant challenges to treating physicians. We report our experience of mortality patterns in this condition over the last 2 decades. There is paucity of literature on this aspect of JDM.

Patients and methods Case files of 72 patients diagnosed to have JDM at a single centre during the period January 1993-November 2012 were reviewed. The total follow-up amounts to 256 patient years. The diagnosis of JDM was based on Bohan and Peter criteria. Standard treatment protocols (based on glucocorticoids and methotrexate) were used in management and the senior author was involved in the care of all these children. We recorded 7 deaths amongst these patients over the last 2 decades.

Results Amongst the 7 children who died, there were 5 boys and 2 girls. The mean age at diagnosis was 10.2 years (range 7–12 years) and duration of treatment ranged from 1 month to 9 years. The causes of death included: upper gastrointestinal vasculitis (3 patients) – of the latter, 1 had perforation peritonitis and 1 had massive upper gastrointestinal bleeding leading to sudden death even before medical aid could be provided; interstitial lung disease with secondary infection (2 patients); bronchiolitis obliterans with organising pneumonia and pneumothorax (1 patient); progressive muscle disease with cardiomyopathy refractory to immunosuppesive therapy (1 patient) – this child died after 9 years of follow-up. Intercurrent and breakthrough infections were recorded amongst 5 of these 7 children.

Conclusion JDM is associated with significant mortality – 13.8% at our centre over the last 2 decades. Gastrointestinal and pulmonary involvement remain the most common causes of death in this condition. Infections are also an important contributory cause. In our experience, delays in diagnosis and referral contribute significantly to mortality in this condition.

G232 USE OF INFLIXIMAB IN CHILDREN WITH REFRACTORY KAWASAKI DISEASE

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Introduction The aetiology of Kawasaki diasese (KD) is still an enigma. It is known that cytokines have a major role to play in the pathogenesis of this condition. There is evidence of TNF-α

activation during the acute phase of KD. Although the drug of choice for treatment of KD is still intravenous immunoglobulin (IVIg), approximately 10–15% patients may not respond to the first dose of IVIg. In such cases, an additional dose of IVIg can be given. Infliximab, a TNF-α inhibitor, is also being increasingly used in such situations. We present our experience with use of infliximab in 9 patients with refractory KD at a tertiary care cente.

Patients and methods We diagnosed 112 children with KD during the period January 2007 – November 2012. The diagnosis of KD was based on the American Heart Association criteria. All children underwent detailed echocardiography examinations by a paediatric cardiologist. Standard treatment protocols (IVIg 2 g/kg along with aspirin) were used in the management of these children. In addition, 9 children with KD received infliximab (REMICADE) during this period. The drug was used in doses of 5–7 mg/kg and given intravenously,. Indicationas for using infliximab were as follows: refractory KD in 6 children; 3 children had come in late (>3 weeks) and had coronary artery abnormalities (CAA) at presentation. 7 patients had developed CAA abnormalities before infliximab therapy: transient mild dilatation-2; aneurysms-3; giant aneurysms-2.

Results Complete response to therapy with cessation of fever occurred in 7/9 patients. Platelet count and C-reactive protein concentrations decreased following infliximab infusion in all, except 2. The latter 2 patients, on follow-up, progressed and developed features consistent with systemic onset juvenile idiopathic arthritis. 4 children with mild CAA (dilatation/small aneurysms) showed complete resolution on follow-up; 2 children with giant aneurysms showed no further progression. No side-effects were noted and there were no significant infusion reactions.

Conclusion Infliximab may have a role in refractory KD, especially when there are CAA. In older children, the use of infliximab is also more economical than IVIg.This has therapeutic implications in resource constrained settings of a developoing country.

G233 AORTIC WALL STIFFNESS STUDIES IN CHILDREN WITH KAWASAKI DISEASE WITHOUT CORONARY ARTERY ABNORMALITIES – A FOLLOW-UP STUDY FROM NORTH INDIA

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Introduction The main focus of concern in Kawasaki disease (KD) hitherto has been on children who develop coronary artery abnormalities (CAA). There is emerging evidence to suggest that several long term cardiovascular sequelae (e.g. endothelial dysfunction), can occur in children with KD even without CAA. Abdominal aortic wall stiffness (AWS) is a surrogate marker of endothelial dysfunction. We evaluated AWS in a cohort of 30 North Indian children with KD without CAA, at least 5 years after the acute phase. There is paucity of literature on follow-up studies of AWS in KD in this setting.

Patients and methods 30 children with KD without CAA, diagnosed at least 5 years back, and who had a normal echocardiography examination at time of enrolment were included in the study. The diagnosis of KD was made on basis of American Heart Association criteria. Age, sex and socio-economic status matched healthy children were included as controls. All children underwent abdominal aorta studies by an experienced cardiologist (RM). Clinical measurements were obtained on Siemens Acuson Sequoia C512 Echocardiography Machine. The study protocol was approved by Institute Ethics Committee.

Results Mean age of cases was 11.24±3.48 years with a mean interval of 7.72 ± 2.37 years from initial diagnosis of KD. Mean aortic strain was 0.345 ± 0.145 in cases as against 0.369 ± 0.123 in controls ($p > 0.05$). Mean pressure strain elastic modulus (SEM) was 124.454 ± 57.052 N/m^2 in cases as against 112.526 ± 48.752 in controls ($p > 0.05$). Mean normalised pressure SEM was 1.898 ± 1.023 in cases as against 1.700 ± 0.700 in controls ($p > 0.05$). Mean peak flow velocity was 0.931 ± 0.277 m/sec in cases and 0.915 ± 0.215 in controls. No statistically significant differences could be detected between cases and controls in terms of aortic root diameters.

Conclusion Though a distinct trend towards higher AWS and lower distensibility was discernible in children with KD (without CAA), the difference did not achieve statistical significance. This could be because of the small sample size in this study. More long-term studies on a greater number of subjects are needed to ascertain the extent of endothelial dysfunction in children with KD without CAA. This may have implications on long-term prognostication of children with KD without CAA.

G234 ACUTE TRANSVERSE MYELITIS AS THE INITIAL MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS IN A CHILD

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Introduction Systemic Lupus Erthematosus (SLE) is a chronic autoimmune disease characterised by multisystem involvement. It is complicated by neurological manifestations in 25–95% of the patients. Acute transverse myelitis may be a complication in 1–2% of patients but in some it may even be the initial manifestation of SLE.

Case report 13 years old previously healthy girl was admitted with a history of pain in her lower limbs for 2 weeks, fever for 2 days and sensory loss. She developed acute flaccid quadriparesis over the next 48hrs, with bilateral weakness of her lower limbs, urinary retention and sensory loss from T2 spinal segment down for light touch and pain. Her vibratory and proprioception were absent in her lower limbs. Power in her upper limbs was reduced. She had no cranial nerve involvement and her ophthalmology assessment was unremarkable. She did not have bulbar involvement or respiratory difficulties. Investigations revealed raised inflammatory markers and CSF showed pleocytosis with an elevated protein. She didn't fulfil the ACR criteria for SLE but her blood results were consistent for an active SLE (ANA positive – speckled pattern, double stranded DNA, Anti Sm and Anti RNP antibody positive with low complement C3 and C4 level). Her cultures for blood, urine, CSF were negative as well as extensive virology screen and serology for Lyme's disease. MRI Spine showed high signal within the spinal cord from C5 down to the conus suggestive of multiregional multifocal transverse myelitis. Her MRI brain was normal. She was treated with 7 cycles of cyclophosphamide and methylprednisolone and plasmapharesis. She was wheelchair bound but has responded well to the treatment and is able to walk few steps without support, manage with crutches only to walk and only uses wheelchair occasionally. She doesn't have any upper limb problems at the moment although still has neuropathic bladder.

Conclusion It is important to include autoimmune screen as part of transverse myelitis work up as SLE may not present with typical clinical features at the outset.

Early diagnosis and aggressive treatment of transverse myelitis is important in SLE to prevent long-term disability.