**Introduction:** There is no internationally agreed definition of ‘JDM-Scleroderma overlap’ in children and little data on the clinical characteristics, laboratory features and outcome in this specific group of JDM. The goal of this survey is to achieve consensus on a meaningful definition of JDM-Scleroderma overlap in paediatric population and to determine whether overlap features affect outcome and management.

**Objectives:** To develop criteria for the diagnosis of JDM-Scleroderma overlap using an international consensus process.

**Methods:** A survey was circulated to all investigators in the Juvenile Dermatomyositis Cohort and Biomarker Study and Repository (JDCBS) and extended to other members of the Network for JDM in Paediatric Rheumatology European Society (PReRS JDM working party) through a Delphi survey. Each individual was asked to identify those clinical manifestations that were felt to be most characteristic to enable them to make the diagnosis of JDM-Scleroderma overlap. The opinions on the importance of identification this subtype of JDM were recorded and the role of myositic autoantibodies has been mentioned in the questionnaire.

**Results:** The survey had a response rate of 26.9% (41 individuals) from both JDCBS and PReRS JDM working party. The most common clinical features identified by survey responders as found in JDM patients with scleroderma overlap were sclerodactyly, sclerodermatous skin change proximal to MCP joints, Raynaud phenomenon and ulceration at the tip of fingers, respectively. 95.1% of responders thought the scleroderma overlap presentations influence the outcome of JDM, while 86.4% agreed that these features influence the choice of treatment. Interestingly, all of responders (100%) thought that negative myositic autoantibodies could not exclude the diagnosis JDM-Scleroderma overlap, but almost 11% used positive myositic autoantibodies to diagnose this subgroup of JDM. Myositis associated autoantibodies occurring in systemic sclerosis overlap (anti PM-Scl, anti U1RNP and anti KU) were thought to be the most commonly associated autoantibodies with JDM-Scleroderma overlap (78%), followed by systemic sclerosis specific autoantibodies (anti Sc170, anti RNA polymerase III) (44%). Although 83% felt that autoantibody profile influences the outcome, only 49% thought that autoantibody profile would influence the choice of medication. 89% of participants in this survey have seen patients with JDM-Scleroderma overlap.

**Conclusion:** This process identified clinical features that clinicians felt to be helpful or important in the diagnosis of JDM-Scleroderma overlap. A further process of secondary survey is necessary to agree an internationally acceptable, clinically useful set of classification criteria.

**Disclosure of Interest** None Declared

**P023**

**THE USE OF INTERLEUKIN 1 RECEPTOR ANTAGONIST (ANAKINRA) IN KAWASAKI DISEASE: A RETROSPECTIVE CASE SERIES**

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**Introduction:** Persistent fever and inflammation after infusion of 2g/kg of IVIG, the standard treatment of KD represents a high-risk situation for coronary aneurysms in Kawasaki disease. Identifying patients at risk for IVIG resistance is difficult outside the Asian population, and there remains a critical unmet need to identify an anti-inflammatory treatment that is efficacious in all KD patients. Recent evidence from studies in animals and humans suggest a critical role for interleukin-1 (IL-1) α and β in the pathogenesis of KD.

**Objectives:** To identify the clinical characteristics, reasons for use and response to treatment with anakinra in a retrospective series of patients with Kawasaki Disease (KD).

**Methods:** A retrospective chart review of patients treated with anakinra for KD diagnosed according to the AHA criteria. We compared clinical, biological and echocardiographic characteristics of KD before and after anakinra use. We analysed reasons for use of anakinra, and compared treatment regimens used in 7 European KD referral centres.

**Results:** Eight boys and 3 girls with treatment-refractory KD, aged 4 months to 9 years old, received at least 2 different KD treatments prior to anakinra, which was given on mean at 25 days after disease onset (8 to 87 days). The main reasons for use of anakinra were clinical and biological inflammation, progression of coronary dilatations, and severe myocarditis or cardiac failure. Doses of anakinra ranged

**Disclosure of Interest** None Declared
from 2 to 8 mg/kg and duration varied from 6 to 81 days. On anakinra treatment, fever disappeared within hours (<24 h) in 3 patients, and within 2 and 6 days in two patients respectively. Six others patients were not febrile at onset of anakinra. In addition, CRP levels fell two to three fold within 48 hours in 7/9 evaluable patients. In terms of its effect on coronary artery dilatation, Z scores decreased in 10/11 patients and increased in one who died suddenly of pericardial hemorrhage.

Conclusion: Anakinra used late in the disease course led to a rapid and sustained improvement in clinical and biological inflammation. However, our retrospective analysis did show neither a striking nor a rapid decrease of coronary dilatations and we cannot determine if anakinra itself had an effect on coronary artery dimensions. More robust data will be available soon from the two Phase II ongoing trials, KAWAKINRA using anakinra treatment early after one failure of IVIG treatment (European Clinical Trials no, 2014-002721-41) and ANAKID (ClinicalTrials.gov identifier: NCT02179853), focused on patients with coronary giant aneurysms.

Disclosure of Interest
None Declared

PO24
OUTCOME OF 24 PATIENTS WITH CHILDHOOD-ONSET TAKAYASU ARTERITIS: A RETROSPECTIVE STUDY IN TWO TERTIARY CENTRES IN JAPAN
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Introduction: Takayasu arteritis (TA) is a chronic large-vessel vasculitis affecting the aorta and its major branches. Glucocorticoids (GCs) can achieve remission in most patients with TA. As most patients suffer relapse after reducing GCs, additional therapy such as methotrexate, azathioprine and intravenous cyclophosphamide (IVCY) has been applied. Recently, the efficacy of TNF-α inhibitors or tocilizumab (TCZ2) has been tested. However, there remain insufficient data on clinical characteristics and outcomes of patients with childhood-onset TA, including recent therapeutic advances.

Objectives: We aimed to retrospectively analyse the characteristics and outcomes of patients with childhood-onset TA in two tertiary centres in Japan.

Methods: Subjects were 24 patients with TA diagnosed when younger than 18 years. Follow-up duration was more than 2 years. Diagnosis of TA was based on the EULAR/PRINTO/PRES criteria. We divided the subjects into two groups; those who did and did not experience relapse. Relapse was defined as NIH criteria for active disease in TA.

We analysed the patients’ background, treatment and complications. We additionally compared two groups according to prednisolone (PSL) reduction rate and whether or not they carried HLA-B52.

Binary data were analysed by Fisher’s exact test, and quantitative data by Mann-Whitney U test. The cumulative relapse rates after the diagnosis were estimated using the Kaplan-Meier method; differences between two groups were assessed by the log-rank test.

Results: Median age at onset was 13.0 years (range: 0.5-18.0), years old (males:female, 11:13). Thirteen patients (54%) had HLA-B52. Period from onset to diagnosis was 1 (0-66) months. Angiographic classifications were I, II, III, IV and V (n=3, 2, 4, 1, 2 and12), respectively. Four patients and one patient had pulmonary artery involvement and coronary involvement, respectively.

Three patients achieved drug-free remission. Median dose of prednisolone (PSL) was 0.8 (0.4-2.4) mg/kg/day at initiation of treatment and 0 (0-0.3) mg/kg/day at the last visit. Immunosuppressants were used in 13 (54%) from the onset and in 21 (88%) at the last visit. IVCY was used in 10 patients (42%) from the onset, and 4 after the relapse.

Some biologics were used in 4 (17%) from the onset, and in 12 (50%) at the last visit.

Twelve patients (50%) experienced relapse, and median number of relapses was 3 (1-6) times. The relapse rate was not significantly different between the HLA-B52 positive and negative patients (69% vs 27%, P=0.099). However, median duration from diagnosis to the first relapse was significantly shorter in HLA-B52 positive patients than negative patients (6 vs 51 months, P=0.016). The reduction rate of PSL was significantly greater in relapsed patients than non-relapsed patients (0.058 vs 0.025 mg/kg/month, P=0.017). The cumulative relapse rates after the started IVCY or TCZ were not significantly different between the patient who introduced IVCY (n=14) and TCZ (n=12) (P=0.10).

Nine patients (38%) had cardiovascular complications (aortic valve regurgitation (AR) (n=4), anulvalaortic ectasia (AAE) (n=1) and AR+AAE (n=4)). Two patients underwent cardiac surgery. Ten patients had osteoporosis, 2 had surgery for glaucoma and 5 had some mental disorders in their clinical outcomes. Our study suggests the outcome of childhood-onset TA is unfavourable.

Disclosure of Interest
None Declared

PO25
MYOSITIS DAMAGE INDEX IN A COHORT OF INDIAN CHILDREN WITH JUVENILE DERMATOMYOSITIS
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Introduction: International Myositis Assessment and Clinical Studies Group (IMACS) developed the Myositis Damage Index (MDI) structured separately for pediatrics and adults, to provide consistency in myositis outcomes. The MDI documents persistent changes in 11 organ systems thought to be related to damage. There is a dearth of studies on damage caused by Juvenile Dermatomyositis (JDM) especially from less resource countries like India where our study.

Objectives: Primarily to assess the myositis damage index in a cohort of children with JDM from a single centre in Mumbai, India and secondarily to study associations of factors leading to long term damage in these children.

Methods: After ethics approval and consents, a total of 23 patients with JDM under regular treatment and at least 2 year follow up, at first study visit were identified. Specifically excluded were children with overlap syndromes eg scleromyositis. The MDI was assessed as severity of damage and extent of damage at the first study visit and reassessed at a second visit at least six months later and only damage present at both visits were scored to give a final severity and extent of damage score. There were no drop outs.

Results: 23 children with age range at disease onset 1y - 17.9 y (mean 6.9 y, median 6.8 y, IQR 3.5-8.8) were diagnosed as JDM after a duration of symptoms ranging 1m-30 m (mean 6.8 m, median 5 m, IQR 3.8-5.5). They age at the study visit ranged from 5.5 y - 24.8 y (mean 13.4 y, median 12.9 y, IQR 10.4-15.3) after a follow up duration ranging from 2 y - 20.1 y (mean 5.9 y, median 4.2 y, IQR 2.6-8.5, Total 136.5 patient years). The disease course was mononcylic in 14, continuous in 6 and polyoncylic in 3. The total MDI extent of damage score ranged from 0- 8 / 35 (children) or 37 (adolescents) (mean 2.04, median 2.0, IQR 0.5-2.5) and severity of damage score ranged from 0- 24.7 /710 (mean 4.7, median 3.5, IQR 1.3-6.4). 17/23 children had damage in one or more organ systems, with 9 showing damage in one organ system, 4 in two organ systems, 3 in three organ systems.