being 5 months. Mean duration of disease at the time of enrolment was 7.18yrs. Disease course was monocyclic in 24 (68.6%). Muscle strength was normal in 71.4%. Severe involvement defined as MMT8 score below 64 was seen in 8.6%. Cutaneous activity was determined by aCAT with 40% children having some form of cutaneous activity. Based on MYOACT, 31.4% children had evidence of disease activity at the time of cross-sectional assessment with skin being the commonest organ system involved in 28.6% followed by muscles in 22.9%. Twenty-one (60%) children had some form of cutaneous damage. Calcinosis in 12 (34.3%) and lipodystrophy in 8 (22.9%). Twenty four subjects had an MDI score of $\geq$1 suggesting damage in at least one organ system. Most commonly affected organs were skin, endocrine and muscles in 20, 12 and 9 subjects respectively. Nine (25.7%) subjects in our study had some form of a physical dysfunction suggested by a CHAQ score above 0. Previous studies on long-term outcomes in children with JDM have either not used validated outcome measures or have used fewer measures [1–3].

Conclusions: Highlight of our study is the use of validated outcome measures for evaluation of long-term outcomes. After mean disease duration of 7.18 yrs, 1/3rd subjects had evidence of disease activity with almost 1/10th having moderate to severe activity. About 2/3rd had damage in at least one organ system. Skin was the most common organ affected by activity as well as damage. About 1/4th had reduced physical functioning. Thus, JDM is not a disease where one time treatment would suffice and regular long-term follow-up is required. Counselling of the caregivers is also critical for them to adhere to follow-up. Larger long-term studies using validated outcome measures are required to confirm these findings.

References:

[1] Singh S, Bansal A. Twelve years experience of juvenile dermatomyositis in North India. Rheumatol. Int. 2006;26:510–5.
[2] Huber AM, Lang B, LeBlanc CM, Birdi N, Bolaria RK, Malleson P, et al. Medium- and long-term functional outcomes in a multicenter cohort of children with juvenile dermatomyositis. Arthritis Rheum. 2000;43:541–9.
[3] Ravelli A, Trail L, Ferrari C, Ruperto N, Pistorio A, Pilkington C, et al. Long-term outcome and prognostic factors of juvenile dermatomyositis: a multinational, multicenter study of 490 patients. Arthritis Care Res. 2010;62:63–72.

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THU0494 CLINICAL FEATURES OF CHILDREN WITH KAWASAKI DISEASE IN DIFFERENT AGE GROUPS IN SOUTHWEST CHINA

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Background: Kawasaki disease (KD) is a systemic vasculitis characterized by acute and prolonged fever. The prevalence of coronary artery abnormality (CAA) is as high as 11%. The young infants usually have the increased risk of CAA, but do not have the typical clinical manefestations of KD.

Objectives: To explore clinical features of children with KD in different age groups to improve the prognosis of KD.

Methods: A total of 218 children with kawasaki disease were divided into the infants group, the toddlers' group, the pre-school age group and the school age group. Retrospective analysis of clinical data were performed among the groups. Categorical data were compared with each other statistically by Chi-square analysis. Statistical significant was defined as $P < 0.05$. Due to the insufficient cases of school age group and five cases of patients with entire clinical data, the analysis was focused on the other three groups and excluded the five cases in the following statistical analysis.

Results: (1) Among the 218 KD patients, the male to female ratio was 1.5:1 and the recurrence rate was 1.8%. Seven cases (3.2%) were diagnosed as atypical KD, and 84 (38.5%) patients accepted intravenous gamma globulin (IVIG) treatment after the sixth day of KD onset. The incidence of IVIG-resistent KD was 8.7% and the rate of coronary dilation was 11.5%. (2) Fever was the most common clinical feature (100%). The bilateral bulbar conjunctiva injection and the change in mucosa of oropharynx were 85.4% and 81.2% respectively. Moreover, cough (40.5%), diarrhea (16.9%) and vomiting (8.5%) were also very common in the present KD patients. (3) Patients from the toddlers' group were more common to develop lymphadenopathy and skin rash (χ^2=7.784, P=0.02; χ^2=10.794, P=0.005), but were less frequently to be documented with cough and diarrhea (χ^2=7.334, P=0.026; χ^2=18.447, P=0.000). (4) The incidence of increased platelets was more common in the infants group (χ^2=7.552, P=0.023). Comparing with the urine test among three groups, the toddlers' group had a higher incidence of sterile pyuria (χ^2=10.653, P=0.005), and infants younger than 12 months old had a lower incidence of proteinuria and positive urine ketone (χ^2=15.507, P=0.000; χ^2=40.336, P=0.000).

Conclusions: The respiratory tract, the digestive and urinary systems are involved commonly in Kawasaki disease, and patients from different age groups showed different clinical features, which should be pay more attention to promote the prognosis.

References:

[1] Kowalczyk M,Turska-Kmiec A,Ziolkowska L,et al. [Symptoms, diagnosis and characteristic abnormalities in the coronary arteries in Kawasaki disease in children]. Med Wieku Rozwoj, 2010. 14(4):344–349.
[2] Burns JC,Shike H,Gordon JB,et al. Sequelae of Kawasaki disease in adolescents and young adults. J Am Coll Cardiol, 1996. 28(1):253–257.
[3] Newburger JW,Takahashi M,Gerber MA,et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Circulation, 2004. 110(17):2747–2771.DOI: 10.1161/01.CIR.0000145143.19711.78.

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THU0495 EFFECTIVENESS OF CHILDHOOD VACCINATIONS IN CAPS PATIENTS TREATED WITH CANAKINUMAB: RESULTS FROM AN OPEN-LABEL PHASE 3 EXTENSION STUDY

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Background: Canakinumab (CAN) has been shown not to impair antibody production following vaccination in children in an open-label phase 3 study (NCT01302860).[1] Here we present the results from the extension of this study.

Objectives: To evaluate the presence of protective antibody levels following immunisation with inactivated vaccines in CAPS patients during extension study.

Methods: Patients who completed the core study were allowed to continue into the extension study on the standard dosing regimen of 2 mg/kg subcutaneous CAN every 8 weeks or on last dose/dosing regimen received in the core study. Vaccination response was evaluated using post-vaccination antibody titres at 4 and 8 weeks after immunisation. Patients were considered assessable for an antibody response to a specific vaccination if they had a measurement of antibody titre 0–14 days post-vaccination (pre-vaccination assessment) and at least 1 subsequent measurement of antibody titre at 4 weeks and/or 8 weeks post-vaccination. However, for patients with adequate pre-dose antibody titres and maintained during the trial, the specific patient vaccination was deemed non-assessable.

Results: During the extension phase, of 17 patients ($\leq$6 years), 4 received 8 types of vaccinations against *Corynebacterium diphtheriae*, *Bordetella pertussis*, *Neisseria meningitidis*, *Clostridium tetani*, influenza type A and type B, *Haemophilus influenzae* B, *Streptococcus pneumoniae*, or hepatitis B. Of 20 unique patient-vaccination cases, 17 were assessable for a vaccination response, whereas for the remaining 3, pre-dose antibody titre was not available. For 16 (94.1%) assessable cases, post-vaccination antibody titres increased above protective levels. For one patient who received Tetravec formulation (diphtheria, tetanus and acellular pertussis combination), the response observed for 1 (vaccination against *Clostridium tetani*) of the 3 vaccines included in Tetravac represented optical density rather than antibody concentrations and hence considered non-evaluable. For 19/20 patient-vaccinations, including those without pre-dose antibody titres, protective levels were observed during the study, which were maintained throughout the extension study.

Conclusions: Canakinumab appeared to have no effect on post-vaccination antibody production following the administration of non-live vaccines in CAPS patients.

References:

[1] Brogan P, et al. Arthritis Rheumatol. 2015;67:(S10).

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THU0496 PULMONARY SYMPTOMS AS THE FIRST PRESENTATION OF KAWASAKI DISEASE IN CHILDREN

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Background: Kawasaki disease (KD) is a medium vessel vasculitis which predominantly affects children less than 5 years of age. Though principal clinical features are mucocutaneous, KD in children may have multiple systemic manifestations, including pulmonary, which may create diagnostic difficulties for the treating physician.

Objectives: We describe our experience of managing children with uncommon pulmonary presentation of KD.