Kawasaki disease (KD) or mucocutaneous lymph node syndrome is a systemic vasculitis in children that involves medium-sized vessels with predilection for coronary arteries. Due to the high probability of cardiovascular complications, an early diagnosis and treatment is required. Objective: To describe demographic, clinical and analytical features of Kawasaki disease in children and their families.

Background: Kawasaki disease (KD) or "mucocutaneous lymph node syndrome" is a systemic vasculitis in children that involve medium-sized vessels with predilection for coronary arteries. Due to the high probability of cardiovascular complications, an early diagnosis and treatment is required. Objective: To describe demographic, clinical and analytical features of Kawasaki disease in children and their families.

Methods: We set up an observational study of patients with KD in a University Hospital between Jan-94 and Dec-18. Classic classification criteria were used for diagnosis. Diagnosis of aneurysms was made by echocardiography, echocardiography and ergometric test (performed 39/53 pts). Conversely, all 13 pts with CAE showed a normal cardiac visit, whilst ECG was abnormal in 1 patient (7.1%).

Conclusion: Our long-term follow-up in a large, even monocentric, cohort reports possible risk factor of CAL according to current literature. Our long-term follow-up assesses, in real life, the benign course of KS in children without CAL after 6-8 weeks from onset. According to recent guidelines, stopping cardiologic assessment in no risk pts results economically advantageous, timesaving and able to reduce emotional discomfort in children and their families.

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Table 1.

<table>
<thead>
<tr>
<th></th>
<th>No-CAL</th>
<th>CE</th>
<th>CAE</th>
</tr>
</thead>
<tbody>
<tr>
<td>N pts</td>
<td>288</td>
<td>58</td>
<td>15</td>
</tr>
<tr>
<td>Median age at disease onset</td>
<td>2 y 1 m</td>
<td>2 y 5</td>
<td>5 m</td>
</tr>
<tr>
<td>Median duration of fever (days)</td>
<td>7</td>
<td>8</td>
<td>11</td>
</tr>
<tr>
<td>Average value of CRP (mg/dl)</td>
<td>6.81</td>
<td>8.38</td>
<td>14.15</td>
</tr>
<tr>
<td>Day of first dose of IVIG</td>
<td>8</td>
<td>7</td>
<td>11</td>
</tr>
</tbody>
</table>

Table 2.
Disclosure of Interests: José Luis Martín-Varillas: None declared, D. Prieto-Peña: None declared, Lara Sánchez Bilbao: None declared, Eva Peña Sainz-Pardo: None declared, Belén Atienza-Mateo: None declared, Monica Calderón-Goercke: None declared, Itirgo González-Mazón: None declared, Natalia Palmou-Fontana: None declared, Maria Teresa Viadero Ubierna: None declared, María Jesús Cabero: None declared, Miguel A González-Gay Grant/research support from: Prof. MA González-Gay received grants/research supports from Abbvie, MSD, Jansen and Roche., Speakers bureau: Consultation/fees/participation in company sponsored speaker’s bureau from Pfizer, Lilly, Sob, Cellgen, Novartis, Roche and Sanofi., Ricardo Blanco Grant/research support from: Abbvie, MSD, and Roche, Consultant for: Abbvie, Pfizer, Roche, Bristol-Myers, Janssen, Speakers bureau: Abbvie, Pfizer, Roche, Bristol-Myers, Janssen.

REFERENCES

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SAT0508 AN INTERNATIONAL SURVEY ON APPROACHES TOWARDS IMMUNISATION IN CHILDREN WITH RHEUMATIC DISEASES: A REPORT OF THE PRES VACCINATIONS WORKING GROUP

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Background: Data on immunisation practices in Paediatric Rheumatology are scarce. EULAR recommendations for vaccination in paediatric patients with rheumatic disease (RD) were published in 2011. In some countries national recommendations for vaccination of immunocompromised patients are available.

Objectives: To ascertain the opinion and current practices of paediatric rheumatologists with regards to immunisation of children with rheumatic diseases (RD), and to establish their confidence to immunise the patients with RD on immunosuppressive medication with live vaccines in light of the evidence available.

Methods: An online survey of practices and opinions towards immunisations with a focus on immunisation of the immunocompromised child with RD was distributed to paediatric rheumatologists across the globe. Responses were collected via SurveyMonkey and descriptive analysis was performed. Responses were anonymous with the exception of identification of country and length of practice.

Results: 289 responses were received from 53 countries in Europe, North and South America, Australia and Asia. 35% of the respondents had over 15 years of practice in Paediatric Rheumatology, while 42% had 5 - 15 years. 57% responded that all immunisations or at least part of them are given in their paediatric rheumatology unit, and 60% that the vaccinations are mandatory in their country. 93% of respondents support the immunisation of paediatric patients with RD, 6.9% responded that they are either not supportive/not sure/support only vaccinations with inactivated vaccines. 53% reported that national recommendations for immunisations of immunosuppressed child are available but not specific to Paediatric Rheumatology. 41% of respondents inform their practice on immunisation of patients with RD based exclusively on the EULAR recommendations, 37.5% based on national guidelines, 8.5% on local guidelines and 10% on combinations of the above. 48% of clinicians would postpone vaccinations in all cases if disease is active.

In terms of immunisations with live vaccines of patients with JIA on immunosuppressive treatment, 41% of respondents would recommend the first dose of MMR or Varicella vaccines to patients with stable disease on Prednisolone < 1 mg/kg/day (maximum 20 mg) for less than 1 month or higher dose up to 2 mg/kg/day for less than 14 days, 14% would also recommend these vaccines if the above steroid dose was given in combination with Methotrexate (MTX) < 15mg/kg/week, 30% would recommend these vaccines if the patient was on MTX monotherapy. Compara- ble percentages reported confidence to also recommend booster doses of the two vaccines for the above drug combinations (45%, 15.7%, 37%, respectively), whilst up 10% of respondents would recommend them to patients on anti-TNF agent alone, and up to 7% for other biologics. For patients with SLE and quiescent disease on similar medications as above, 41%, 15%, 31%, respectively, of clinicians reported confidence to recommend MMR or Varicella booster doses.

48% of the respondents identified the reluctance of other health professionals involved in the process of immunisations as the main reason hampering the vaccination of paediatric patients with RD, whilst 22% indicated parental refusal or hesitancy.

Conclusion: There is variation in practice and opinions worldwide with regards to immunisations in paediatric patients with RD, and this likely reflects the discrepancies between national guidelines for immunisation of immunosuppressed child and also national policies. More studies are required, but there is an increasing vote of confidence towards immunisation of patients on lower grade immunosuppression with MMR or varicella vaccines.

Disclosure of Interests: None declared


SAT0507 EVALUATION OF THE NEW CLASSIFICATION CRITERIA FOR PFAPA SYNDROME

Fabio Crimi1, Manel Mejbri2, Véronique Hentgen3, Glory Dingulu2, Isabelle Koné-Paul2, Sophie Georgin-Lavialle4, Pascal Pillet4, Michael Hofer5, JIRcohorte. Roche, Consultant for: Abbvie, Pfizer, Roche, Bristol-Myers, Janssen, Ricardo Blanco Grant/research support from: Abbvie, MSD, and Roche, Consultant for: Abbvie, Pfizer, Roche, Bristol-Myers, Janssen, Ubierna: None declared, María Jesús Cabero: None declared, Miguel A Martinez-Varillas: None declared, Monica Calderón-Goercke: None declared, Itirgo González-Mazón: None declared.

Background: Modified Marshall criteria used for PFAPA syndrome have never been validated and are little used by the experts because the symptoms of monogenic fevers often overlap with PFAPA ones. A new set of classification criteria based on an international survey and a consensus conference in Genoa was developed in 2018.

Objectives: Evaluate the performance of the new criteria in a real-life setting.

Methods: This is a multicentric, prospective and descriptive cohort study, through the recurrent fever module of the JIRcohorte platform. 417 patients diagnosed with PFAPA (187), monogenic fever syndromes (167) or unclassified recurrent fever syndrome (UPF=63) from Swiss and French centers were enrolled in the study. The new classification criteria were applied to this cohort and we calculated their performance. We then analyzed which of the criteria performed the less well.

Results: One hundred fourteen from 187 (61%) PFAPA patients met the new criteria, as well as 20/230 non-PFAPA patients (FMF: 3, MKD: 4, ankylosing spondylitis: 1, skin rash: 10, other: 7). The criteria showed a good specificity but an insufficient sensitivity. Excluding the skin rash criterion among PFAPA patients not meeting the criteria was done among PFAPA patients not meeting the criteria was useful, with a sensitivity of 80% which could be a fair compromise for PFAPA classification criteria.

Conclusion: Genoa 2017 classification criteria for PFAPA syndrome showed a good specificity but an insufficient sensitivity. Excluding the skin rash criterion among PFAPA patients not meeting the criteria was useful, with a sensitivity of 80% which could be a fair compromise for PFAPA classification criteria. Our study highlights the difficulty in establishing classification criteria due to the lack of gold standard for PFAPA diagnosis.