IMMUNOLOGICAL FINDINGS AND RESPIRATORY JOINT EFFUSIONS: AN INDICATOR OF UNFAVOURABLE OUTCOME OF CRMO IN CHILDREN

Joanna Tsigkouli, 1Benjamin Jacobs, 2Cristina Ilea, 2Alessandro Vidoni. 1Whittington Hospital; 2Royal National Orthopaedic Hospital

10.1136/archdischild-2021-rcpch.506

Background Chronic Recurrent Multifocal Osteomyelitis (CRMO) is an autoinflammatory, chronic disease that usually appears in childhood. It can be unifocal or multifocal, and main symptoms are pain and swelling on the area of the affected bones. Most of the times, it has a benign and self-limiting course; however, there is a minority of patients for whom the disease remits often and the symptoms are not well limiting course; however, there is a minority of patients for whom the disease remits often and the symptoms are not well

Abstract 1244 Table 1 Comparison of average HbA1c(mmol/mol) pre- and post-lockdown grouped by baseline HbA1c

<table>
<thead>
<tr>
<th>Group</th>
<th>Number of Subjects</th>
<th>Pre-LD (1)</th>
<th>Post-LD (2)</th>
<th>Av-Post-LD (3)</th>
<th>Improved 1–2</th>
<th>Improved 1–3</th>
<th>Improved 2–3</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;69</td>
<td>38</td>
<td>86.67</td>
<td>76.94</td>
<td>76.49</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>64–69</td>
<td>25</td>
<td>65.69</td>
<td>61.82</td>
<td>61.07</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>58–63</td>
<td>44</td>
<td>60.14</td>
<td>58.31</td>
<td>59.78</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>53–57</td>
<td>23</td>
<td>55.35</td>
<td>54.23</td>
<td>54.63</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>48–52</td>
<td>18</td>
<td>50.51</td>
<td>52.25</td>
<td>52.59</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>&lt;48</td>
<td>13</td>
<td>42.07</td>
<td>44.24</td>
<td>46.50</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
</tbody>
</table>

The inclusion criteria were all dates, languages and ages; humans; and clinical relevance. The exclusion criteria were: no reference to A-T, not an original article, animal studies, not clinically relevant.

Results Search returned 194,890 articles; 14,622 titles and abstracts were reviewed after removing 180,268 duplicates.

Full text review of 1,163 articles was performed and 1,039 studies were included (13,459 exclusions, 124 excluded after full text review).

The most frequently reported immunoglobulin abnormality was low IgA in the classical group, followed by high IgM. The majority of immunoglobulin results reported in the variant group were normal. The most likely abnormality in the variant group was high IgM. Very limited age data were reported.

Other common immunoglobulin abnormalities in the classical group included low IgG2 (n=283), low IgG3 (n=44) and low IgG4 (n=86).

699 cases in the classical group received replacement immunoglobulin, starting at a median age (n=40) of 4 years 6 months (range 2 months to 16 years 0 months, IQR 3 years 0 months to 8 years 0 months). No cases in the variant group were reported to be on replacement immunoglobulin. 230 cases in the classical group and 3 in the variant group were reported to have received prophylactic antibiotics.

Recurrent infections were reported in 1118 cases in the classical group and 18 in the variant group. The most frequently reported recurrent infection was sinusopulmonary infection in the classical group (n=570) and lower respiratory tract infection/pneumonia in the variant group (n=11).

Non-infectious respiratory manifestations reported in the classical group included bronchiectasis (n=228), interstitial lung disease (n=35), unspecified chronic lung disease (n=3), pulmonary fibrosis (n=7), and pneumothorax (n=50). Age data (where available) will be reported on these manifestations. No non-infectious respiratory manifestations were reported in the variant group.

Conclusions There is a wide variety of infectious and non-infectious respiratory manifestations, and immunological abnormalities reported in classical A-T. There are fewer immunological and respiratory manifestations reported in variant A-T.

We are grateful to Action for A-T, the A-T Society, and BrAshA-T, for financial support.
controlled, while others even develop other autoimmune diseases, such as arthritis and sacroiliitis.

Objectives To determine if remote joint effusions seen on whole-body MRI can predict non-response to Pamidronate and/or unfavourable outcome in paediatric CRMO patients.

Methods A total of 81 cases were retrospectively reviewed. The patients who had joint effusions on MRI which had no adjacent CRMO lesion were compared to patients with poor response to Pamidronate treatment and to the ones with poor outcome (severe pain resistant to medication and/or new autoimmune disease). Joint effusions that had an adjacent CRMO lesion were seen as reactive and the patients were added to the non-effusion group.

Results Most patients in this cohort were females (n=59, 73%) and most common lesion was the clavicle (n=35, 43%). 14 of the patients (17%) had remote joint effusions. Partial or no response to Pamidronate was seen in 15 patients. 5 patients (6%) have severe uncontrollable pain, 7 (9%) have arthritis, 1 has psoriatic arthritis, and 2 (2%) have sacroiliitis. Comparisons among these three characteristics (remote joint effusions, poor response to Pamidronate, poor outcome) were statistically significant (p<0.001), showing that these three characteristics usually co-exist. Furthermore, this group of patients did not have other features of SAPHO (Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis) syndrome, in which synovitis and CRMO can co-exist. Positive family history was not related to any of these three characteristics.

Conclusions There is a group of CRMO patients who have a distinct, more active disease course than the rest of the patients. When joint effusions with no NBO lesion is identified on MRI, it should be taken into account prognostically and thus avoid bisphosphonate treatment. Further research is needed to determine if these patients respond better to other treatments such as disease-modifying anti-rheumatic drugs (DMARDs) or TNF inhibitors.

Paediatric Educators’ Special Interest Group

1249 TEACH PAEDIATRICS AND INSPIRE: USING QIP METHODOLOGY IN EDUCATION PROMOTES PROSPECTIVE TRAINEES’ INTEREST IN A CAREER IN PAEDIATRICS

Benjamin Rosen, Jonathan Pass, Karishma Desai. The Royal Free NHS Trust

Background Recruitment of paediatric trainees is not matching its increasing demand. In 2019, only 69.8% of paediatric training program posts were filled, the lowest out of all UK programmes. Such workforce shortages cause burnout, which negatively impacts patient care. Positive experiences drive medical students’ choice of that specialty, yet for all of 2019, at Barnet hospital (BGH) and Royal Free Hospital (RFH), over a quarter of students did not even score their paediatrics placement as ‘satisfactory’.

Objectives Use QIP methodology to improve Paediatric teaching across the trust to achieve an increase of self-reported likelihood of medical students to choose paediatrics of ≥20%, by the end of their placement.

Methods During the 2-month audit phase we engaged the departments and medical school at all levels with consultation, fishbone diagrams and presentations.

The following measures were used

- Outcome: Percentage change in students’ likelihood to pursue a career in paediatrics. A comparison of their self-reported likelihood, as recorded before and after placements.

Quality Improvement and Patient Safety

1248 PERCEPTION OF PPE (PERSONAL PROTECTIVE EQUIPMENT) AMONGST PAEDIATRICIANS

Pramod Nair, Yazhiri Kodeeswaran, Nisrien Eltag Mohamed Osman, Satanupa Banerjee. Bedford Hospital NHS Trust

10.1136/archdischild-2021-rcpch.507

Background PPE (Personal Protective equipment) use has been mandatory due to the current pandemic with Covid-19 and has been in use for the past 1 year. Use of PPE in paediatrics comes with its own challenges but is likely to be used more frequently in future. At this juncture it would be useful to collect feedback from the paediatricians to improve our understanding of the practical issues with use of PPE so that as a group changes or alterations could be considered if at all necessary.

Objectives To understand practical difficulties with use of PPE if any and review personal experience from paediatricians who have been using them in the current pandemic

Methods Survey regarding PPE specific questions were sent to multiple Paediatricians and the results analysed

Results 96 paediatricians responded to the survey. Of the 96 doctors who responded, 61 were consultants and 35 were at a middle grade level. 27% of the respondents said wearing face masks did not affect their interactions with children. 73% of the respondents noted difficulties with increased stranger anxiety, difficulties with communication and establishing rapport with their patients. Of the 73% who had difficulties with interaction this was highest in the 1–3 year patient age group. 61% of the respondents did not feel that having face masks interferes with handovers although 39% felt it does. Almost 80% of the respondents felt that having full PPE interfered with their procedural skills with particular difficulties with intubation (33%) and cannulation (56%). 54% respondents preferred goggles and 46% preferred visor for eye protection. 30% of the respondents said that they would still prefer to continue to use PPE even after the pandemic.

Conclusions This survey brings out some very interesting facts (and many other comments) about use of PPE in our professional life. A large majority felt that it affects their interactions with children particularly the 1–3 year age group due to the difficulty in reading expressions and communication. It would be important to look at having alternative face masks which are child friendly. A significant proportion felt that having full PPE interferes with their procedural skills and it is important that this is urgently addressed as it could have significant patient safety implications. Fogging, poor visual clarity etc. were many of the issues reported and the wider paediatric group needs to escalate these concerns so that alternatives could be considered. PPE is likely to remain a long term piece of equipment to be used in paediatrics and it is important that we now ensure that they are tailored based on feedback from its users.