

and every 6 months from 2014 to 2017, through a standardized auto-questionnaire.

Results: 278 patients (M/F: 0.29) aged 12.7 years on average (\pm 4.9, range 2-21) were taken care of between 2014 and 2017 by RESRIP. Juvenile idiopathic arthritis (JIA) ($n = 142$), connective tissue disease ($n = 49$) and auto-inflammatory disease (AID) ($n = 32$) are the 3 main pathologies covered. Among the 278 patients, twenty-one percent of patients needed academic support when entering the network, including: 37% in the JIA group, 34% in the connective tissue group and 18% in the AID group. Educational assistance was set up for all patients with school difficulties at inclusion but also for all the patients during their follow-up.

Patient Support: 178 Individual Action Plan (IAP) were implemented by RESRIP. Twelve patients benefited from additional time for their school exams and 10 were allowed to pass the baccalaureat (French final college exam) over two years instead of a year. In thirty patients, sports education has been adapted. Forty-one MDPH files (Departmental Houses for the Disabled) were produced to enable the establishment of a school life assistant (AVS), teaching materials and/or technical aids. Finally, 8 patients received home schooling assistance through Home Learning Assistance Services (SAPAD).

Education professional support: RERSIP established partnerships with the French National Education (FNE) and the SAPAD (home tuition service for ill children). For the FNE, RESRIP provided school doctors or nurses with: 8 continuing medical training and 25 personal interviews to explain the pathologies. In addition, 5 multidisciplinary meetings, within the institutions were organized, for 5 patients with social integration difficulties and stigmatization. RESRIP actions, has allowed a significant decrease in school absenteeism between 2014 and 2017: 3.2 days per year on average at day 1 to 0.5 days per year on average ($p < 0.05$) at 42 months.

Conclusion: Over time, a better understanding of the impact of chronic illness over school and education professionals, has allowed RERSIP to improve its support. The result is a notable decline in school absenteeism and in unjustified physical education exemptions. Several projects are underway: The development of a standard IAP available for doctors, the set-up of a partnership with hospital school, willing to help for the implementation of home school support courses in addition to SAPAD.

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THU0536 LONG TERM IMMUNITY IN A PAEDIATRIC COHORT OF PATIENTS WITH RHEUMATIC DISEASES FROM A TERTIARY HOSPITAL IN SPAIN

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Background: Pediatric patients with rheumatic diseases (RD) are at increased risk of infections. Vaccines have been proved to be very effective to prevent them. Nevertheless the long-term immunity after vaccination remains quite unknown in this group.

Objectives: To compare the long term seroprotection in pediatric patients with rheumatic diseases who received measles, rubella, mumps, tetanus, diphtheria, hepatitis B, Hib and meningococcus C vaccination according to the routine immunization schedule in Spain, and healthy children.

Methods: We designed a cross-sectional study including consecutive pediatric patients with RD who attended the rheumatology clinic and healthy children older than 10 y.o. The administered vaccines, treatment and pathology of each patient will be recorded. Their antibodies titers against each antigen were quantified and compared to healthy children.

Results: 60 patients (median age 13 y.o IQR 10.2-12.7) and 15 healthy children (mean age 11.3 y.o. IQR 11.3-12.7) were included. In the patients group 62% were female, and 35% in healthy. Diagnosis: 85% Idiopathic juvenile arthritis, 18% Lupus or juvenile dermatomyositis. 46% had received biologic treatment sometime. Seroprotection rate was (patient vs control): measles 85% vs 81%, rubella 78.8% vs 73.3%, parotiditis 84.7% vs 66.7%; VHB 27% vs 10.5%, diphtheria 89.5% vs 81%, tetanos 64.9% vs 61.1%. Hib 42% vs 53%, pneumococo 92.4% vs 100%, meningococcus C 12% vs 11%. $p > 0.05$

Conclusion: No statistical differences were detected, although the scarce number of control subjects might have influenced this result. There is a tendency towards a lower antibody persistence anti-Hib in patients compared to healthy children. The response to life virus vaccine and tetanus seem to be as good as in healthy children.

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THU0537 ANGIOMATOID FIBROUS HISTIOCYTOMA MIMICKING SYSTEMIC JIA VIA MUTATION-DRIVEN IL-6 PRODUCTION

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Background: Angiomatoid Fibrous Histiocytoma mimicking Systemic JIA via mutation-driven IL-6 production: Angiomatoid Fibrous Histiocytoma (AFH) is a rare tumour associated with mutation-driven production of Interleukin-6 (IL-6) which causes a systemic inflammatory picture similar to Systemic Juvenile Idiopathic Arthritis (sJIA).

Objectives: A previously well 6-year old girl was referred with 6 weeks of abdominal pain, nausea, weight loss, night sweats, & lethargy.

Examination was unremarkable apart from a 2cm lump in the right popliteal fossa. This had been reported on USS 2 months earlier, at an external hospital, to be a Sebaceous cyst. There was no rash, organomegaly, lymphadenopathy, or synovitis.

She had persistent recurrent fevers of $>39^{\circ}\text{C}$, but not in the classical quotidian pattern of sJIA.

Methods: Bloods showed persistent Hb <70 , platelets >600 , ESR >100 , & CRP >200 .

Autoantibody & full infection screens were negative.

Urine HMMA:creatinine ratio was minimally raised at 7.2 (normal 1.8 - 5)

Faecal calprotectin, upper & lower GI endoscopy were normal.

Bone-Marrow Aspiration was reported as being highly reactive but with no malignancy seen.

Whole-body STIR MRI was reported as normal, but repeat localised USS & MRI of the knee showed a 27x18x21mm well circumscribed, mixed cystic/solid lesion with marked vascularity. The lesion was then biopsied, and subsequently excised

Plasma IL-6 levels were significantly elevated at 46.7pg/ml (normal range: 0-2), but normalised after excision. TNF and IL1b levels were normal

Results: Initial biopsy and FISH analysis confirmed the diagnosis of AFH with an abnormal EWSR1 signal pattern and classical EWSR1-CREB1 fusion. She went on to have a full excision, and staging investigations were negative.

AFH is rare ($<0.3\%$ of all soft-tissue tumours) and arises in the deep dermis/subcutis of the extremities of children & young adults. It has been regularly described in the axilla & popliteal fossa. It has a high (15%) local recurrence rate but rarely metastasises.

In $>90\%$ of cases AFH is associated with a characteristic translocation: t(2:22)(q33;q12). This forms the fusion gene EWSR1-CREB1 which in turn leads to continuous activation of CREB1. The promoter region of IL-6 has a CREB1 binding site thus causing IL-6 over-production and leading to the paraneoplastic syndrome.

Conclusion: Paraneoplastic immune dysregulation is a rare, but important cause of symptoms often otherwise associated with rheumatic diseases. Careful physical examination, and liaison with oncology teams, is needed, particularly before treatment with steroids or IL-6 inhibitors such as Tocilizumab are considered

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