Objectives: To describe a cohort of infants and young children with resistant Kawasaki Disease (rKD) who were noted to have prolonged difficult courses with resultant CAA and ultimately diagnosed with Aortitis.

Methods: Demographic, clinical manifestations and imaging data were collected from children seen in Rheumatology clinic with a diagnosis of rKD and aortitis. Nonparametric descriptive statistics were used to describe the data.

Results: Between 2010-2018, 63 out of 417 KD referrals were diagnosed with rKD. 7 children had prolonged time to CRP normalisation, prolonged admission or recurrence of symptoms (Table 2). All patients underwent Magnetic Resonance Angiography (MRA) and were found to have evidence of large vessel arteritis consisting of wall irregularity, thickness and contrast enhancement.

Table 1

ID	Age at diagnosis (mo)	Number of clinical signs**	BCG site inflammation	Lowest Hemoglobin g/DL	Highest WBC 10 (9)/L	Highest Platelet 10 (9)/L	Highest CRP mg/L	Lowest Albumin g/L	Maximum ALT U/L	Maximum AST U/L
A	31	3	0	NA	NA	NA	NA	NA	NA	NA
B	7	4	0	9.5	37.3	875.0	206.0	21.0	36.0	32.0
C	4	4	1	9.8	43.2	918.0	130.0	22.0	44.0	52.0
D	3.5	4	1	9.2	42.4	1715.0	181.4	18.0	61.0	99.0
E	5	4	1	6.7	17.6	1413.0	148.2	27.0	20.0	24.0
F	2	3	0	6.4	35.6	959.0	212.1	15.0	8.0	19.0
G	24	NA	NA	8.6	21.9	1002.0	181.4	27.0	17.0	30.0

5 patients were male; All were Singaporean Chinese except for G who was from the Philippines; A & G were treated in an outside facility/separate service initially and some data was not available; # Blood test results were generally taken at the same time apart from the platelet count; **Only D presented with lymphadenitis.

Table 2

ID	Time to CRP normalization (weeks)	Number of IVIG given	Steroids	Time to abnormal echo (weeks)	Time to aneurysm development (weeks)	Other outcomes
A	*	2	oral only	4	4	1 st IVIG given 3 weeks after symptom onset
B	*	2	IV/PO	2	2	deceased, MI secondary to thrombus
C	2	1	IV/PO	1.1	0	treated with Infliximab and then Methotrexate,
D	5	1	IV/PO	0.3	2	post infliximab psoriasis, treated with Infliximab and then Methotrexate, multiple infections in hospital; BCG reactivation while on Infliximab
E	9	2	no	1.4	2.7	UTI, AIHA, lost to follow up
F	3	3	IV/PO	0.71	2.5	post Infliximab psoriasis, multiple infections in hospital
G	7	2	oral only	7	0	lost to follow up

*patients A & B had recurrent KD episodes interspersed with short periods of symptom resolution

Conclusion: While transthoracic echocardiograms remain imaging investigation of choice, for children with recalcitrant or recrudescing KD, especially in the setting of delayed CRP resolution of >2 weeks; an MRA should be considered.

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THU0509

IMPACT OF PEDIATRIC RHEUMATIC DISEASES ON MEXICAN CAREGIVERS.

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Background: Pediatric rheumatic diseases (PRD) have an important impact on different aspects of the patients' and caregivers' life, such as physical, emotional, economic, and social. Some studies have shown that parents of patients with PRD have important impact but there is a lack of information of this topic from Latinamerican countries.

Objectives: The aim of this study is to describe and analyze the impact of juvenile idiopathic arthritis (JIA), juvenile systemic lupus erythematosus (JSLE), and juvenile dermatomyositis (JDM) on Mexican primary caregivers.

Methods: This is a multicenter cross-sectional study conducted in third-level reference pediatric hospitals in Mexico from December 2018 to November 2019. We included primary caregivers of pediatric patients with JIA, JSLE, and JDM that were treated in participant centers.

CAREGIVERS questionnaire, a validated multiassessment tool to measure the impact of PRD on caregivers, was applied to the participants. Collection of social, demographic, and clinical data was also performed and correlated with questionnaire results.

Results: A total of 200 primary caregivers participates in the study (109 JIA, 28 JDM, and 63 JSLE), aged 38 (IQR 32 – 46), mostly women (84.5%), from 6 centers, representing 13/32 Mexican states (Figure). One third (78) had a remunerated job, 123 (61.5%) had a relationship, 77 (38.5%) reached high school or higher, and 131 (65.5%) spends more than one hour to get to the center. Patients cared aged 12 (IQR 9 – 16), mostly women (67%), 87 (43.5%) with active disease, 43 (21.5%) with any disability, 94% and 29% treated with DMARD and biologics, respectively.

Feelings of worry and sadness predominant at diagnosis that decreased over time (42.5% and 28.5% vs 9.5% and 31.5%, respectively) and changed for peace (44%). Concerns about disabilities were more frequent on JIA group (34%), while pain and economic issues in JSLE (47% and 30%, respectively). Most of the caregivers feel anxiety about the future of their patients (148, 74%), regardless of the diagnosis. Participants reported that the way they spend the time, social life, and personal health worsened since diagnosis (49.5%, 32%, and 34.5%, respectively), especially in those with JSLE (60%, 39%, 46%). In 126 (63%) participants the economic situation worsened, 129 (64.5%) borrowed money (76% in JSLE, P = .03), 63 (31.5%) had problems to buy medications, and 48.5% have had problems at work. In 25 (12.5%), the family relationship was affected after diagnosis, however, the main supportive network reported were family members.

Conclusion: This work described the main impacted areas in life of primary caregivers of patients with PRD, showing a perspective of the burden of the disease.

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THU0510

CHOREA AND CARDITIS: AN UNEXPECTED COMBINATION. THE NEW FACE OF ACUTE RHEUMATIC FEVER

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Background: Acute Rheumatic Fever (ARF) is an immunomediated multisystem disease that occurs about 2-5 weeks after Group A *Streptococcus Pyogenes* beta-hemolytic (GAS) pharyngitis. After a negative peak in the 1980s, following the introduction of antibiotic prophylaxis, the disease is currently recovering. Rheumatic carditis is one of the most worrying aspects as it is still one of the major causes of cardiovascular death in the young-adult population. However, if diagnosed early and treated, sequelae with aortic and mitral valve involvement can be prevented.

Objectives: The aim of our study is a description of Acute Rheumatic Fever in all its manifestations in a cohort of pediatric patients belonging to the Azienda Ospedialiera Universitaria Integrata of Verona.

Methods: A retrospective analysis was conducted collecting all the cases of ARF, diagnosed by Jones's criteria, related to Pediatric Rheumatology and Pediatric Cardiology of Verona from January 2005 to December 2019. Demographic and clinical data were collected for all patients such as clinical presentation, disease evolution and cardiac involvement.

Results: 73 patients were analyzed, of whom 53 had an acute onset of ARF and 20 received a diagnosis of previous ARF due to indolent carditis. The prevalent age at the time of diagnosis in both groups was between 5 and 14 years of age. Among patients with acute onset, carditis was the most frequent major manifestation (94.3%), followed by polyarthritis (41.5%), chorea (24.5%) and erythema marginatum (7.5%). Only in one patient we could observe subcutaneous nodules (1.8%). Regarding the minor manifestations, the increase in inflammation markers was present in 83% of cases and fever was present in 75.5%, followed by arthralgia (58.4%) and prolonged of PR interval to ECG (9.4%). Carditis was also present in all 13 patients who presented chorea. Clinically, previously unknown heart murmur occurred in 28 patients. Therefore, the mismatch between cardiac objectivity and carditis finding is clear: infact, compared to an important finding of carditis (50 patients) only slightly more than half of the patients (28 patients) showed an evident clinic finding. Finally, no correlation was found between the levels of the antistreptolysin O titer and the severity of heart damage. Patients with early diagnosis of carditis were treated at onset with corticosteroid therapy according to the American Heart Association scheme and did not show valvular cardiac outcomes. A patient who received a late diagnosis of carditis currently presents a significant and permanent cardiac damage despite adequate steroid treatment undertaken at the time of diagnosis.

Conclusion: The description of this cohort of pediatric patients shows that the ARF has not disappeared in industrialized countries. Treatment of streptococcal infection (primary prophylaxis) plays a key role in preventing ARF. Of great impact is the prevalence of carditis which is present in 94.3% of patients. Early diagnosis is therefore of primary importance and the subsequent follow-up path, consisting of periodic therapy with penicillin (secondary prophylaxis) and periodic cardiological checks, greatly affects children's quality of life. Chorea, unlike what has been described in the literature, occurred simultaneously with the cardiac process, while the cutaneous manifestations (subcutaneous nodules and erythema marginatum), once pathognomonic of the rheumatic disease, are today of rare observation.

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THU0511

THE RISK FACTORS OF HIP OSTEOARTHRITIS IN JIA PATIENTS: THE DATA FROM A SINGLE CENTER RETROSPECTIVE CONTINUOUS STUDY

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Background: Hip osteoarthritis (HOA) is a severe irreversible complication of patients with juvenile idiopathic arthritis (JIA) leads to intensive pain, disability and required total hip arthroplasty (THA) in childhood or in the young adults.

Objectives: the aim of our study was to evaluate risk factors of HOA in JIA patients.

Methods: we analyzed 753 patients with JIA for last 10 years. In each patient we observed if the HOA developed or no. We evaluated the main clinical JIA measurements, such a JIA category, CRP, ESR, WBC, PLT, active joints, initial or delayed hip involvement, ANA, HLA B27, onset age, JIA duration, time before

HOA and before THA, bone metabolic markers, treatment, particularly glucocorticosteroids (GCS), their cumulative doses, route of administration (oral, intra-articular, pulse therapy). HOA was confirmed with radiological assessment (MRI and CT). For comparison analysis used nonparametric statistics. Each possible variable was evaluated with univariate regression analysis and further multiple regression analysis was applied.

Results: the comparative study between 2 groups shown in the table 1. HOA was detected in 48 (6.4%), in oligoarthritis-0%, in polyarthritis - 4.9%, in psoriatic arthritis - 5.0%, in enthesitis-related arthritis -11.8%, in systemic arthritis 19% (p=0.0000001), THA was performed in 16 (2.1%). Hip involvement at onset of JIA was similar in both groups (49.5% vs 41.7%, p=0.367).

Table 1. Characteristics of JIA patients, depends on HOA development.

Parameter	HOA (n=48)	No HOA (n=705)	p
Onset age, y	8.0 (4.0; 12.5)	5.9 (3.0; 10.7)	0.045
HLA B27, n (%)	13/25 (52.0)	88/283 (31.1)	0.033
JIA duration, y	7.5 (3.2; 12.1)	4.2 (1.8; 7.2)	0.00007
CRP, mg/l	2.8 (0.8; 20.9)	1.3 (0; 7.1)	0.006
Active joints, n	11.0 (6.0; 27.0)	6.0 (3.0; 12.0)	0.000001
Oral GCS, n(%)	21 (43.8)	131/704 (18.6)	0.00003
GCS pulse-therapy, n (%)	21 (43.8)	114/702 (16.2)	0.000002
GCS, cumulative doses, mg	5000 (3000; 16000)	2000 (1000; 4000)	0.001
Biologics, n (%)	43 (89.6)	308 (43.7)	0.0000001
Remission, n (%)	24 (50.0)	461 (65.4)	0.03
Time, before remission, y	7.4 (3.9; 12.3)	3.0 (1.4; 6.2)	0.0000001
Alkaline phosphatase, U/l	129.3 (84; 244)	223 (147; 386)	0.00001
Delayed hip involvement, n (%)	28 (58.3)	24/87 (27.6)	0.00004

We calculated cut-off and OR of variables, associated with HOA development (table 2). After selection of 26 clinically meaningful and statistical significant risk factors only 22 pass the univariate regression analysis. In multiple regression analysis the main independent risk factors of HOA development were: HLA b27 (p=0.001), oral GCS/pulse therapy GCS (p=0.03) and alkaline phosphatase≤165 U/l (p=0.00006).

Table 2. Cutt-offs, odds ratio for HOA risk factors in JIA patients.

Parameter	Se	Sp	OR (95%CI)	p
Active joints > 4	89,6	42,0	6,2 (2,4-15,9)	0,000002
Alkaline phosphatase ≤ 165 U/l	65,9	70,8	4,7 (2,4-9,2)	0,000001
GCS > 2700 mg	82,6	58,1	6,6 (2,1-20,6)	0,0004
Time before remission>5 years	66,7	78,9	7,5 (3,1-18,0)	0,0000001
Time before biologics >7,8 years	44,2	86,7	5,2 (2,6-10,2)	0,0000001

Conclusion: the main risk factors of HOA were increased inflammatory activity, systemic arthritis, HLA B27, systemic corticosteroids, delayed remission and biologics, decreased bone metabolism. Delayed hip involvement underline the possible non-inflammatory mechanism of HOA, such as avascular necrosis.

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THU0512

HEALTH RISK BEHAVIORS IN ADOLESCENTS WITH JUVENILE IDIOPATHIC ARTHRITIS (JIA) IN THE COURSE OF DISEASE: RESULTS OF THE GERMAN MULTICENTER INCEPTION COHORT (ICON)

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Background: Health risk behaviors (HRB) are defined as specific forms of behavior associated with increased susceptibility to a specific disease or ill health on the basis of epidemiological or social data. The social implications of HRB include being able to use them as a way to gain respect and acceptance from peers, establishing independence from parents, or providing a subjective feeling of maturity and adequate stress management [1].

Objectives: Since chronically ill adolescents such as those suffering from JIA can develop a HRB that is challenging for optimal care the aim of this study was to compare the frequency of HRB in adolescents with JIA and their peers, and to determine whether they change during the course of disease.

Methods: Patients ≤ 16 years of age with recently diagnosed JIA (< 12 months) were enrolled in the inception cohort of patients with newly diagnosed JIA (ICON), an ongoing prospective observational, controlled multicenter study