Abstract AB1072 Table 1. Clinical features of patients according to the anatomical classification of uveitis.

		Anterioruveitis 21 cases (56.8%)	Intermediateuveitis 12 cases (32.4%)	Posterioruveitis 1 case (2.7%)	Panuveitis 3 cases (8.1%)	Total 37 cases (100%)
Gender	**Female**	9(24.3%)	6 (16.2%)	1(2.7%)	1(2.7%)	17(45.9%)
	Male	12(32.4%)	6(16.2%)	0	2 (5.4%)	20 (54%)
Ocular involvement	**Unilateral**	3 (8.1%)	3(8.1%)	0	1(2.7%)	7 (18.9%)
	Bilateral	18 (48.6%)	9 (24.3%)	1(2.7%)	2(5.4%)	30 (81%)
Etiology	**Idiopathic**	13 (35.1%)	11(29.7%)	0	2(5.4%)	26 (70.2%)
	JIA	5 (13.5%)	0	0	0	5(13.5%)
	BehcetDisease	0	1(2.7%)	0	1(2.7%)	2(5.4%)
	Sarcoidosis	1(2.7%)	0	1(2.7%)	0	2(5.4%)
	TINU	2(5.4%)	0	0	0	2(5.4%)

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AB1072B THE CONSEQUENCES OF THE PROVISIONAL PAEDIATRIC RHEUMATOLOGY INTERNATIONAL TRIALS ORGANISATION JUVENILE IDIOPATHIC ARTHRITIS CLASSIFICATION CRITERIA

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Background Last year the International League of Associations for Rheumatology (ILAR) classification criteria for juvenile idiopathic arthritis (JIA), [1] were challenged by the provisional Paediatric Rheumatology International Trials Organisation (PRINTO) classification criteria.[2] Four disorders were proposed: (a) systemic JIA; (b) rheumatoid factor (RF)-positive JIA; (c) enthesitis/spondylitis-related JIA; and (d) early-onset antinuclear antibody (ANA)-positive JIA. Early-onset ANA-positive JIA is defined by: arthritis for ≥ 6 weeks, and early-onset (≤ 6 yrs), and presence of 2 positive ANA tests with a titer ≥ 1/160 at least 3 months apart with the exclusions of having systemic JIA, RF-positive arthritis, or enthesitis/spondylitis-related JIA.
Objectives To evaluate the shifts from the original subtypes of JIA in the new disorder of early-onset ANA–positive JIA.
Methods This study used data from the international PRINTO based registry regarding pharmacovigilance in JIA called Pharmachild.[3] For this analysis we used the data of 4,165 patients completely categorized following the ILAR 'oligoarthritis', 'RF-negative polyarthritis', 'psoriatic arthritis' and 'undifferentiated JIA' (UJIA) subtypes and with complete determination of ANA status. These patients were if possible reclassified in the early-onset ANA–positive JIA according to the provisional PRINTO classification criteria.
Results Table 1 shows the characteristics of all 4,165 patients according to the ILAR criteria. Of this final set of 4165 patients, 1279 (30.7%) were ANA-positive and 957 (74.8%) classified into the PRINTO 'early onset ANA-positive JIA' category. Of these 957, 2 patients were RF-positive, which is an exclusion criterion for the 'early onset ANA-positive JIA' category and therefore were not categorized as early onset ANA-positive JIA. The female proportion was higher than in any ILAR subtype being 83.0% (793/955). The origin (ILAR categories) of the 955 patients in the 'early onset ANA-positive JIA' category consisted of 33.7% patients with persistent oligoarthritis (322/955), 24.7% (236/955) with extended oligoarthritis, 28.0% with RF-negative polyarthritis (267/955), 4.2% with psoriatic arthritis (40/955) and 9.4% with UJIA (90/955).

	Persistent OJIA	Extended OJIA	RF negative PJIA	Psoriatic arthritis	UJIA
Total number of patients, n (%)	1283 (30.8%)	663 (15.9%)	1665 (40.0%)	210 (5.0%)	344 (8.3%)
Age of disease onset, years (IQR)	4.5 (2.5-8.3)	3.7 (2.3-1.7)	6.7 (2.9-11.3)	8.6 (3.4-13.3)	5.7 (2.6-10.8)
Female, n (%)	968 (77.6%)	541 (81.6%)	1283 (77.1%)	145 (69.0%)	228 (66.3%)
ANA positive, n (%)	424 (33.0%)	293 (44.2%)	384 (23.1%)	51 (24.3%)	127 (26.9%)

Table 1. *Patient characteristics (4165 patients) divided in ILAR 2001 categories; correctly determined (taking all exclusion criteria into account).*
OJIA= oligoarticular JIA, PJIA= polyarticular JIA

Conclusion This study shows that of all ANA-positive JIA patients belonging to the 'oligoarthritis', 'RF-negative polyarthritis', 'psoriatic arthritis' and 'UJIA' ILAR subtypes, 74.8% met the criteria for the PRINTO 'early onset ANA-positive' category. The female proportion was higher than in any ILAR subtype being 83.0%. This new category consists largely of 3 ILAR subtypes: persistent oligoarthritis (34%), extended oligoarthritis (25%) and RF negative polyarthritis (28%). Further studies on these provisional criteria are ongoing.

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Disclosure of Interests Vera Mars: None declared, Joost F. Swart: None declared, Gabriella Giancane: None declared, Sytze De Roock: None declared, Anne Estmann: None declared, Marija Jelusic: None declared, Estefania Moreno Ruzafa: None declared, Jaime de Inocencio: None declared, Jelena Vojinovic: None declared, Agustin Remesal: None declared, M Laday: None declared, Rolando Cimaz: None declared, A V Cochino: None declared, Inmaculada Calvo Grant/research support from: received research grants from Pfizer, Roche, Novartis, Clementia, Sanofi, MSD, BMS and GSK, Consultant for: Advisory boards: Novartis, AbbVie, Speakers bureau: AbbVie, Roche, Novartis, SOBI, M Harjacek: None declared, Nico Wulffraat: None declared, Nicolino Ruperto Grant/research support from: The Gaslini Hospital, where NR works as full-time public employee, has received contributions (> 10.000 USD each) from the following industries in the last 3 years: BMS, Eli-Lilly, GlaxoSmithKline, F Hoffmann-La Roche, Janssen, Novartis, Pfizer, Sobi. This funding has been reinvested for the research activities of the hospital in a fully independent manner, without any commitment with third parties., Consultant for: Received honoraria for consultancies or speaker bureaus (< 10.000 USD each) from the following pharmaceutical companies in the past 3 years: Ablynx, AbbVie, Astrazeneca-Medimmune, Biogen, Boehringer, Bristol-Myers Squibb, Eli-Lilly, EMD Serono, GlaxoSmithKline, Hoffmann-La Roche, Janssen, Merck, Novartis, Pfizer, R-Pharma, SanofiServier, Sinergie, Sobi and Takeda., Speakers bureau: Received honoraria for consultancies or speaker bureaus (< 10.000 USD each) from the following pharmaceutical companies in the past 3 years: Ablynx, AbbVie, Astrazeneca-Medimmune, Biogen, Boehringer, Bristol-Myers Squibb, Eli-Lilly, EMD Serono, GlaxoSmithKline, Hoffmann-La Roche, Janssen, Merck, Novartis, Pfizer, R-Pharma, SanofiServier, Sinergie, Sobi and Takeda.
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AB1072C CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS THE CORRELATION BETWEEN CAROTID INTIMA-MEDIA THICKNESS AND MARKERS OF INFLAMATION TO DETERMINE PRE-CLINICAL ATHEROSCLEROSIS

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Background: Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in childhood. JIA is a heterogeneous group of disorders with different disease progression and prognosis. Cardiovascular

morbidity and mortality are becoming major health concerns for children with inflammatory rheumatic diseases. Inflammation has been suggested to play an important role in the pathogenesis of both atherosclerosis and JIA.
Objectives: objective of this study was to determine carotid intima-media thickness (cIMT) in children with JIA and their correlation with JIA subtype and markers of inflammation.
Methods: We included 112 patients (50 boys and 62 girls) with diagnosis of JIA for at least 6 mouths and 54 healthy control subjects (32 girls and 22 males).These all children were had normal body mass index, blood pressure, lipids and blood glucose levels to exclude conventional risk factors.
Carotid ultrasound for evaluation of cIMT was performed by same radiologist who was blinded to the participants' clinical and laboratory data. Patients datas were collected. We compared all cMIT values of patients and controls and investigated the relationship between inflammatory markers, disease status,periods and therapies.
Results: Our study showed that children with JIA had more increased cIMT values than controls. There was no correlation between disease subgroups, activity status, ANA, HLA-B27, RF positivity, WBC, ESR and CRP with cIMT. The cIMT values were not differences between patients used steroid, DMARD and biologic agents and non used. An increased active disease and total disease periods, decreased in mean platelet volüme were determined as independent risk factors at increased of cIMT.
Conclusion: The study showed that the patients with JIA had more risks than healthy controls for cardiovascular diseases.
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Other orphan diseases

AB1073 A RETROSPECTIVE COHORT STUDY OF IGG-4 RELATED DISEASE IN IRISH PATIENTS

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Background: Immunoglobulin (Ig) G4-related disease (IgG4RD) is a novel clinical entity characterized by elevated serum IgG4 concentration and tumefaction or tissue infiltration by IgG4-positive plasma cells.
Objectives: To describe the clinical presentations, laboratory features, imaging manifestations, histopathologic characteristics and treatments in a cohort of 38 patients with IgG4RD.
Methods: A retrospective study was performed at St. Vincent's University Hospital. Clinical, laboratory, imaging and histopathologic data was retrieved from electronic records. All data were assessed using SPSS 24.0.
Results: Median age was 59 years with M:F ratio= 2.2:1. 24 (63.2%) patients were between 25-65 years), 14 (36.8%) were >65 years. 23 (60.5%) patients fulfilled the Comprehensive Diagnostic Criteria for IgG4RD as 'definite', whereas 5 (13.2%) patients fulfilled 'probable' diagnoses and 10 (26.3%) patients fall in 'possible' category. GI manifestations (followed by pancreatic) were the most frequent clinical presentation. 23 (60.5%) patients presented with single organ involvement; pancreas was the most frequently involved organ (17/38, (44.7%)). 55.3% had a serum IgG4 level above 135mg/dL. Lymphoplasmacytic infiltration was the commonest histopathologic pattern reported in 29 (76.3%) specimens. 25 (65.8%) patients had received steroid therapy and 19 (50.0%) had a good response. 11 (28.9%) patients received immunomodulatory agents including Rituximab (n=4), Azathioprine (n=7), and Mycophenolate mofetil (n=4). Overall, 28 (73.7%) patients had complete remission with treatment.
Conclusion: IgG4RD is a rare entity in Ireland and an inadequately understood condition overall. Further research is required to better understand the pathophysiology, clinical course and optimal treatment for IgG4RD.

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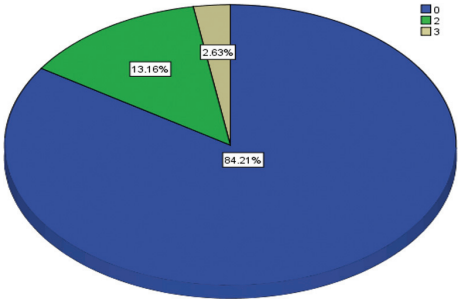
Abstract AB1073 Table 1. Organ involvement in 38 patients with IgG4RD

Organ involved	Number of patients (%)
Pancreas	17 (44.7%)
Bile ducts (Biliary tract)	13 (34.2%)
Retroperitoneal mass	7 (18.4%)
Retro-orbital mass (Orbit adnexa)	4 (10.5%)
Lacrimal glands	3 (7.9%)
Salivary glands	2 (5.2%)
Lungs	2 (5.2%)
Lymph nodes	1 (2.6%)
Thyroid	1 (2.6%)
Testicles	1 (2.6%)
Kidney	1 (2.6%)

Abstract AB1073 Table 2. Histopathologic patterns in 38 patients with IgG4RD

Histopathologic pattern	Number of patients (%)
Lymphoplasmacytic infiltration	29 (76.3%)
Oblitrative phlebitis	2 (5.3%)
Lymphoplasmacytic infiltration + Increased esinophils number	1 (2.6%)
Lymphoplasmacytic infiltration + Storiform fibrosis + Oblitrative phlebitis	1 (2.6%)
Lymphoplasmacytic infiltration + Storiform fibrosis + Phlebitis without obliteration	1 (2.6%)



Abstract AB1073 Figure 1. IGG-4 Responder Index score in 38 patients with IgG4RD

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AB1074 AUTOIMMUNE/INFLAMMATORY SYNDROME INDUCED BY ADJUVANTS—ASIA—RELATED TO BIOMATERIALS: ANALYSIS OF 50 CASES

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Background: Systemic autoimmune or granulomatous disorders related to biomaterials of human use have rarely been described
Objectives: The aim of this study was to report cases of autoimmune/inflammatory syndrome induced by adjuvants (ASIA) related to biomaterial injections and prostheses, mainly silicone, hyaluronic acid, acrylamides and methacrylate compounds in a Spanish patient cohort.
Methods: This study is a retrospective analysis of clinical, laboratory, histopathological and follow-up data of 50 cases of patients suffering from late-onset, non-infectious inflammatory/autoimmune disorders related to bioimplants. Late onset was defined as 3 months or more post injection. Data were obtained through a further non-systematic but comprehensive review of the literature. Fifty cases of late-onset adverse reactions related to biomaterial injections or prostheses were reviewed.
Results: All cases had systemic complaints that could be categorised as ASIA. In all but five patients, inflammatory features at the implantation