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THU0594

SHIARI-JAVADI CRITERIA FOR THE DIAGNOSIS OF GENERALISED JOINT HYPERMOBILITY IN CHILDREN

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Background: Benign joint hypermobility syndrome (BJHS) is the occurrence of musculoskeletal symptoms in hypermobile individuals in the absence of systemic rheumatologic disease.^{1–3} One of the most common criteria for the evaluation of generalised joint hypermobility is Beighton's criteria. However, Beighton criteria were designed for all ages not specifically for children group (according to the children physiological and growth characteristics). Inability to identify limited hypermobility, failure to differentiate generalised joint hypermobility from joint hypermobility syndrome and BJHS and the restrictions on the use of epidemiological studies are further limitations.^{4, 5}

Objectives: Because of the disadvantages of the existing criteria and the high prevalence of generalised joint hypermobility in children, we decided to propose further modifications to Beighton criteria for the diagnosis of joint hypermobility.

Methods: A case-control study was designed with 200 participants from 3 to 16 years of age with 100 children with BJH (according to Beighton criteria) in case group and 100 age-sex matched children as control group. Cases were selected from outpatients Clinic of Rheumatology and the control group was selected from the emergency department of hospital. The case group consisted of children who had musculoskeletal pain complaints or were suspected to be hypermobile. The Beighton criteria were used as the gold standard, and all of cases fulfilled the Beighton criteria. In addition, the participants in the control group should be free of chronic disease or musculoskeletal complaints. For the determination of new criteria we performed literature review on the range of motion in different age and gender paediatric groups, studied the various existing criteria and their differences, and utilised our own clinical observations. Eight manoeuvres were set for examination (Fig 1). All were bilateral, except the neck hyperextension.

Results: There were 42 (42%) male children in each group. The mean age was 6.8 years. Table 2 compares the results of the new and Beighton criteria in both cases and controls. All cases were hypermobile, and two of the 100 controls were hypermobile with the new criteria. Based on these results, new proposed criteria had the sensitivity, specificity, positive predictive value and negative predictive value of 100%, 98%, 100% and 98% respectively.

Conclusions: Shiari-Javadi criteria appears to be useful for detecting hypermobility. In addition, they have also overcome many of the disadvantages of the Beighton criteria, and are easier, more practical and more comprehensive to be used in children. However, further studies are required to validate this criteria.

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THU0595

CLINICAL AND THERAPEUTICAL DIFFERENCES BETWEEN ADULTS AND CHILDREN IN A COLOMBIAN COHORT WITH INFLAMMATORY IDIOPATHIC MYOPATHY

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Background: Age-related autoimmunity is a key differentiating factor with diverse presentations in different age groups possibly depending on immune regulatory mechanisms. Even patients with the same diagnostic category may have different presentations according to age and other factors. Inflammatory Idiopathic Myopathies (IIM) are not the exception with important clinical and prognostic differences between adults and children.

Objectives: To describe the clinical and therapeutic differences between adults and children (younger than 18 years old) in a cohort with IIM

Methods: A cross-section retrospective research was done with data collected between 2014 and 2017 from a population diagnosed with IIM according to Peter and Bohan criteria and followed up for at least six months. Statistical association was evaluated by means of Chi-square and Fisher's tests.

Results: Out of a total of 149 patients, 112 were adults and 37 were children. There was significant difference between the clinical presentation of symmetrical muscle weakness 98% in adults vs 73% in children (p: 0.0001), Gottron's papules 32% vs 89% (p: 0.0001), Heliotrope rash 31% vs 62% (p: 0.0016), Calcinosis cutis 1% vs 37% (p: 0.0001), Dermatological involvement 47% vs 94% (p: 0.0001), Myopathic changes in the electromyogram 41% vs 25% (p: 0.0001), and Myopathy proven biopsy 41% vs 25% (p: 0.0001). Therapeutic options showed a significant difference in the use of steroid therapy 93% vs. 84% (p: 0.045), Methotrexate (58% vs 91% p: 0.0001), Azathioprine (62% vs 27% p: 0.0002) and Chloroquine (31% vs 73% p: 0.0001). Other therapies like Cyclophosphamide (19% vs 5% p: 0.04) and Immunoglobulins (23% vs 5% p: 0.01) were significantly different between adults and children.

Abstract THU0595 – Table 1. Clinical characteristics of Colombian Patients with Inflammatory Idiopathic Myopath (Adults and Children)

	Juvenile n=37		Adults n=112		p-value
	N	%	N	%	
Female	28	75,7	76	67,9	0,42
Clinical characteristics					
Symmetrical muscle weakness	27	73	110	98,2	<,0001
Gottron's papules	33	89,2	49	32,9	<,0001
Heliotrope rash	23	62,2	35	31,3	0,0016
Calcinosis cutis	14	37,8	2	1,8	<,0001
Dermatological involvement	35	94,6	53	47,3	<,0001
Myopathic changes (EMG)	9/23	39,1	68/81	84,0	<,0001
Biopsy-proven myopathy (Positive)	4/16	25	46/54	41,1	<,0001
ANA(+)	14/27	51,9	65/89	73,0	0,0580

EMG, electromyogram

Conclusions: Several differences in the clinical and therapeutic characteristics were found between adults and children with IIM. Adults had more frequently symmetrical weakness, myopathic changes in EMG and biopsy proven myopathy. In contrast children had higher skin involvement. Calcinosis constituted an important manifestation of IIM in children. These results suggest that paediatric IIM is a distinct sub-phenotype related to the early age at onset and possibly mediated by different immune interplay key factors.

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THU0596

MEASUREMENT OF SERUM CALPROTECTIN MRP8/14 IN PAEDIATRIC PATIENTS DIAGNOSED WITH JUVENILE IDIOPATHIC ARTHRITIS AND AUTOINFLAMMATORY DISEASES

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Background: There is evidence of the correlation between serum levels of calprotectin MRP8/14 and disease activity in Juvenile Idiopathic Arthritis (JIA), but the same correlation with other rheumatic diseases in children such as autoinflammatory diseases has not been studied as much.

Objectives: To analyse calprotectin MRP8/14 in serum of patients with juvenile idiopathic arthritis (JIA) and various autoinflammatory diseases with different degrees of activity. To check if there is a correlation between serum MRP8/14 and disease activity.

Methods: Study in two phases: 1) Initial, transversal, in JIA and/or autoinflammatory disease patients, determining serum MRP8/14 (by ELISA test) and collecting clinical data: number of active joints, Visual Analogue Scale parents/patients (VASp), VAS physician (VASph); and analytical: C-Reactive Protein (CRP) and white cells blood count (WBC) (among others). 2) Second prospective phase, repeating the same analysis 3–6 months later. Disease activity was assessed by Juvenile Arthritis Disease Activity Score (JADAS) in AIJ; and by VASp, VASph and CRP in autoinflammatory diseases.

Results: The sample included 90 patients (25 males) between the ages of 1 and 21 years (median 11.1); 61 diagnosed with JIA (excluding systemic category), and 29 with different autoinflammatory diseases (including systemic JIA). 69 patients (20 males, with median age 11.2, 48 AIJ, 21 autoinflammatory) participated in the prospective phase. The main results are shown in table 1.

MRP=myeloid related proteins. JADAS=Juvenile arthritis disease activity score. ACJ=active joints count. VASm=visual analogue scale–physician. VASp=visual analogue scale–parents/patient. CRP=C reactive Protein.

Conclusions: In patients with non-systemic JIA, a positive correlation was found between serum MRP8/14 and JADAS, VASph, VASp, CRP, WBC, neutrophilia and (inversely) haemoglobin.

In autoinflammatory diseases positive correlation has been found with VASm, CRP, leukocytosis, neutrophilia, thrombocytosis and (inversely) haemoglobin. MRP8/14 seems to behave as a marker of activity in both JIA and autoinflammatory diseases, although with a stronger association in the latter.

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THU0597

PULMONARY MANIFESTATIONS IN MIXED CONNECTIVE TISSUE DISEASE WITH JUVENILE AND ADULT ONSET – ARE THERE ANY DIFFERENCES?

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Background: Mixed connective tissue disease (MCTD) presents in childhood in 7%–23% of cases, but there is limited knowledge about the comparability of juvenile and adult onset of the disease. A common and serious manifestation is interstitial lung disease (ILD), possibly more common in adult MCTD according to a retrospective report.¹

Objectives: To compare disease variables in juvenile and adult onset MCTD, particularly regarding pulmonary manifestations, after long-term follow-up.

Methods: Two cohorts consisting of, respectively, 52 patients with juvenile onset MCTD and 90 patients with adult onset MCTD from all regions of Norway were compared. Inclusion criteria were fulfilment of the Kasukawa- or the Alarcon-Segovia criteria. Patients with onset of symptoms before the age of 18 years were considered to have juvenile onset. All patients were clinically examined, including high resolution CT and pulmonary function tests.

Results: Mean age at examination was 28.0 (SD 10.3) in juvenile onset, and 54.3 (SD 13.0) in adult onset MCTD (p<0.001). Age at disease onset was 14.4 (SD4.4) and 37.9 (SD 15.1) years, respectively. Patient and disease characteristics are shown in table 1.

	Juvenile onset	Adult onset	p-value
Male gender, n (%)	8 (15)	28 (24)	0.192
Disease duration from symptom onset, mean (SD)	16.2 (10.3)	17.0 (8.9)	0.766
Anti-RNP levels, median (IQR)	180 (36.0–240.0)	44.0 (7.0–237.0)	0.002
ESR, median (IQR)	8.0 (5.0 to 16.0)	11.0 (7.0–18.5)	0.048
CRP, median (IQR)	0.7 (0.5–2.5)	2.0 (0.7–4.9)	<0.001
ILD, n (%)	14 (27)	39 (43)	0.051
ILD% of TLV, median (IQR)	4.0 (1.0–8.3)	5.0 (1.8–20.0)	0.258
ILD in >20% of TLV, n (%)	1 (2)	10 (11)	0.048
DLCO% pred, mean (SD)	73.0 (12.5)	72.0 (19.4)	0.598
FVC% pred, mean (SD)	89.2 (14.7)	89.2 (20.8)	0.711
FEV1% pred, mean (SD)	89.4 (13.7)	81.2 (20.0)	0.016

Juvenile patients had higher levels of anti-RNP, but lower ESR and CRP compared to adult onset patients. ILD was found in 27% of juvenile and 43% of adult patients (p=0.051), and more adult onset patients had ILD in >20% of total lung volume (TLV). Adult onset patients had lower forced expiratory volume in 1 s (FEV1), but similar diffusing capacity (DLCO) and forced vital capacity (FVC) as the juvenile patients.

Conclusions: ILD tended to be more frequent in patients with adult onset MCTD than in those with juvenile onset, although not statistically significant. More patients with adult onset had affection of >20% of TLV. Juvenile and adult patients

	JIA (n=61)		Autoinflammatory diseases (n=29)	
	1 st determination (n=61)	2nd determination (n=48)	1 st determination (n=29)	2nd determination (n=21)
JADAS	r=0.270* p=0.035	r=0.054 p=0.717		
ACJ	r=0.053 p=0.685	r=0.042 p=0.779		
VASph	r=0.308* p=0.016	r=0.041 p=0.784	r=0.304 p=0.108	r=0.622** p=0.003
VASp	r=0.329* p=0.010	r=-0.023 p=0.877	r=0.341 p=0.082	r=0.341 p=0.141
CRP	r=0.212 p=0.107	r=0.349* p=0.018	r=0.579** p=0.001	r=0.102 p=0.688
WBC	r=0.296* p=0.022	r=-0.071 p=0.633	r=0.749** p=0.000	r=0.693** p=0.000
Neutrophiles	r=0.389* p=0.002	r=0.421** p=0.002	r=0.281 p=0.140	r=0.710** p=0.000
Haemoglobin	r=-0.255* p=0.049	r=-0.023 p=0.828	r=-0.454* p=0.013	r=-0.552** p=0.009
Platelets	r=-0.033 p=0.802	r=0.001 p=0.993	r=0.659** p=0.000	r=0.208 p=0.366