LETTERS TO THE EDITOR

Portuguese referrals to pediatric rheumatology – multicentric study

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Dear editor,

Pediatric Rheumatology (PR) specializes in infantile rheumatic diseases. American[1] and European[2,3,4] referrals to PR revealed delays concerning waiting times/distance, compromising disease outcomes. The Portuguese reality is unknown; thus, we aimed to characterize its pattern of PR referral.

We designed a retrospective, multicenter study, including pediatric patients with rheumatic pathology with a first PR appointment from 01/01/2021-31/12/2021. Patients without inflammatory or other miscellaneous diseases requiring follow-up were excluded. Sociodemographic data, diagnosis and referral characteristics were recorded (referring specialty, timing since symptom onset and number of previous specialties until PR contact). Descriptive and comparative analysis were performed using SPSSv25 and t-test, Mann-Whitney, Kruskall Wallis, Chi-squared and Fisher tests, appropriately.

We amounted 160 patients from five hospitals (median age 11.5 years [IQR 8]; 68.8% female). The most commonly referred pathology was juvenile idiopathic arthritis (JIA-53.8%), followed by autoinflammatory syndromes (13.8%). The commonest referring specialties were Pediatrics (51.9%), General Practice (GP-26.9%) and Orthopedics (8.1%). The first PR appointment occurred within one month of symptoms in 21.9% of patients, after 1-3 months in 27.5%, in 11.9% at 3-6 months, 10.6% at 6-12 months and in 28.1% after one year. Of the latter, 33.3% and 35.6% were referred by pediatricians and GP, respectively. Before contacting PR, 73.1%, 23.1% and 3.8% of the children were seen by 1, 2 and 3 other specialties, respectively. Table 1 depicts this data. Adult Rheumatology associated with a PR referral of dermatomyositis patients, Periodic Fever, Aphtous stomatitis, Pharyngitis and Adenitis (PFAPA) and overlap syndromes (p<0.001); Orthopedics to JIA referral (p<0.001); GP to familial Mediterranean fever (FMF) and undifferentiated connective tissue diseases (UCTD) referral (p<0.001); Dermatology and Nephrology to systemic lupus erythematous (SLE) referral (p<0.001); and Hematology to Behçet's disease and other vasculitis referral (p<0.001). Kawasaki's disease and SLE were consulted within one month of symptoms (p=0.017); FMF after one year of symptoms (p=0.017); JIA after 1-3 months of symptoms (p=0.017); and overlap syndrome, other vasculitis, chronic multifocal recurrent osteomyelitis and PFAPA after 3-12 months of symptoms (p=0.017). The earliest children evaluated in PR (<1 month of symptoms) were referred by Pediatrics (p=0.049), with JIA, SLE and Kawasaki's diagnosis, and the latest by Gastroenterology (p=0.049). Among JIA patients, mean age was 11.4±5.00, and 67.4% were female; 57% were referred by Pediatrics, 23.3% by GP and 15.1% by Orthopedics. Fifty-six percent were evaluated in the first 3 months of symptoms, and 23.5% after one year of symptoms. Pediatricians more frequently referred to PR within one month of symptoms (p=0.014).

Overall, referral to PR was mainly done by Pediatrics, GP and Orthopedics, and JIA was the most common disease, matching its prevalence. Approximately, 1/4 of patients reached PR after more than one year of symptoms, 1/2 in the first three months of symptoms, and 3/4 were previously consulted by only one other specialty. Inactive disease states, as in some JIA subgroups, might delay referrals. SLE and Kawasaki's disease met PR in the first month of symptoms. The earliest referral was by Pediatrics. GP might be bypassing JIA referrals to Pediatrics. Moreover, perhaps due to FMF's idiosyncrasies, GP may find it more flaunting to engage and refer; also the small sample might be inflating this association. Barriers to PR referrals are to be reckoned: 1) uneven national coverage, with a clustering in city centers; 2) long distance to travel, ensuing this last thought, and consolidating PR referral's alternatives; 3) insufficient PR education, leading to lack of confidence in musculoskeletal examination and dropping the required index of suspicion for PR conditions.

As around 25% of patients take more than one year since disease onset to reach PR, further research

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TABLE I. DATA ON THE REFERRED PATIENTS TO PEDIATRIC RHEUMATOLOGY, BRANCHED BY HOSPITAL

	All hospitals N=160)	ULSAM (N=13)	CHLO (N=25)	CHLN (N=60)	CHMA (N=50)	HGO (N=12)	P value
Age in years, median (IQR)	11.5 (3)	12.0 (7)	12.0 (4)	11.0 (7)	11.0 (9)	11.0 (6)	0.376
Gender, n/N (%)							0.892
Male	50 (31.3)	4 (30.8)	8 (32)	21 (35)	13 (26)	4 (33)	
Female	110 (68.7)	9 (69.2)	17 (68)	39 (65)	37 (74)	8 (67)	
Rheumatic disease, n (%)							**0.00**
Juvenile idiopathic arthritis	86 (53.8)	9 (69.2)	10 (40)	**25 (41.7)***	**36 (76)***	6 (50)	***-2.4, 3.1**
Reactive/post-infectious arthritis	2 (1.3)	0	**2 (12)**	0	0	0	
Dermatomyositis	2 (1.3)	**1 (7.7)**	0	1 (1.7)	0	0	
Behçet's disease	9 (5.6)	1 (7.7)	0	3 (5)	3 (6)	2 (16.7)	
Kawasaki Disease	2 (5.6)	0	0	0	**2 (4)**	0	
UCTD	7 (4.4)	0	**6 (24)**	1 (1.7)	0	0	
IgG4-related disease	1 (0.6)	0	0	1 (1.7)	0	0	
Systemic sclerosis	1 (0.6)	0	0	1 (1.7)	0	0	
Autoinflammatory syndromes	22 (13.8)	2 (15,4)	2 (8)	7 (11,7)	7 (14)	4 (33,3)	
Systemic lupus erythematosus	13 (8.1)	0	4 (16)	8 (13.3)	1 (2)	0	
IgA vasculitis	3 (1.9)	0	0	1 (1.7)	1 (2)	1 (8.3)	
Sarcoidosis	1 (0.6)	0	0	1 (1.7)	0	0	
Overlap syndrome	1 (0.6)	0	0	1 (1.7)	0	0	
Other vasculitis	1 (0.6)	0	0	1 (1.7)	0	0	
Miscellaneous conditions	9 (5.6)	0	0	**9 (15)**	0	0	
Referring Specialty, n (%)							**<0.001**
Dermatology	2 (1.3)	0	1 (4)	1 (1.7)	0	0	
Gastroenterology	1 (0.6)	0	0	1 (1.7)	0	0	
Medical Genetics	4 (2.5)	0	0	**4 (6.7)**	0	0	
Hematology	3 (1.9)	0	0	**3 (5)**	0	0	
Infectious Diseases	1 (0.6)	0	0	1 (1.7)	0	0	
General Practice	43 (26.9)	**9 (69.2)***	**12 (48)***	16 (26.7)	**2 (4)***	4 (33.3)	***3.6, 2.6, -4.4**
Nephrology	1 (0.6)	0	0	1 (1.7)	0	0	
Ophthalmology	2 (1.3)	0	1 (4)	1 (1.7)	0	0	
Orthopedics	13 (8.1)	2 (15.4)	0	**1 (1.7)**	**8 (16)**	2 (16.7)	
Otorhinolaryngology	1 (0.6)	0	0	0	1 (2)	0	
Pediatrics	83 (51.9)	**2 (15.4)***	11 (44)	**25 (41.2)***	**39 (78)***	6 (50)	***-2.7, -2.2, 4.6**
Adult Rheumatology	6 (3.8)	0	0	**6 (10)**	0	0	
Time to referral, n (%)							**0.034**
< 1 month	35 (21.9)	1 (7.7)	8 (32)	17 (28.3)	9 (18)	0	
1-3 months	44 (27.5)	3 (23.1)	**2 (8)***	14 (23.3)	**22 (44)***	3 (25)	***-2.4, 3.2**
3-6 months	19 (11.9)	1 (7.7)	5 (20)	5 (8.3)	8 (16)	0	
6-12 months	17 (10.6)	3 (23.1)	1 (4)	8 (13.3)	3 (6)	2 (16.7)	
> 1 year	45 (28.1)	7 (38.5)	9 (36)	16 (26.7)	**8 (16)***	**7 (58.3)***	***-2.3, 2.4**
No. of previous specialties, n (%)							**<0.001**
1	117 (73.1)	9 (69.9)	20 (80)	**58 (96.7)***	**23 (46)**	7 (58.3)	***5.2, -5.2**
2	37 (23.1)	4 (30.8)	4 (16)	**2 (3.3)***	**22 (44)***	5 (41.7)	***-4.6, 4.2**
3	6 (3.8)	0	1 (4)	0	**5 (10)**	0	

* Adjusted residues value – positive association above 1.96 and negative association below -1.96

Abbreviations: CHLN - Centro Hospitalar de Lisboa Norte; CHLO - Centro Hospitalar de Lisboa Ocidental; CHMA - Centro Hospitalar do Médio Ave; IQR - interquartile range; HGO - Hospital Garcia de Horta; NS - not significant; UCTD - Undifferentiated connective tissue disease; ULSAM – Unidade Local de Saúde do Alto Minho

into Portuguese PR referrals' obstacles are imperative, meanwhile raising patients and physicians' PR know-how and awareness. While restructuring medical pathways might demand constitutional appraising, upgrading non-rheumatologists' PR literacy and empowering families and teachers to seek health care in recognition of the first rheumatic symptoms are likely to improve outcomes.

Our study holds limitations: the participation of a low number of centers, not geographically representative of the country's PR distribution; not including JIA subgroups (systemic disease might be referred earlier); not considering waiting times; COVID-19 pandemics' influence on the number/type of referrals.

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