REVISTA BRASILEIRA DE REUMATOLOGIA

www.reumatologia.com.br

SOCIEDADE BRASILEIRA DE REUMATOLOGIA

Original article

Profile of paediatric rheumatology specialists and services in the state of São Paulo

Maria Teresa Terreri[a], Lúcia M. A. Campos[b,], Eunice M. Okuda[c], Clovis A. Silva[b], Silvana B. Sacchetti[c], Roberto Marini[d], Virginia P. Ferriani[e], Maria Heloiza Ventura[f], Taciana Fernandes[g], Juliana O. Sato[h], Elizabeth C. Fernandes[b], Claudio Len[a], Cássia Barbosa[a], Ana Paola Lotito[b], Maria Carolina dos Santos[c], Nádia E. Aikawa[b], Mércia Facó[i], Daniela Piotto[a], Vanessa Bugni[a], Kátia T. Kozu[b], Paulo R. Romanelli[i], Adriana M. E. Sallum[b], Marília Febronio[b], Melissa Fraga[a], Cláudia S. Magalhães[h]*

[a] *Unit of Paediatric Rheumatology, Department of Paediatrics, Universidade Federal de São Paulo, São Paulo, SP, Brazil*
[b] *Unit of Paediatric Rheumatology, Instituto da Criança, Hospital das Clínicas, Faculdade de Medicina, Universidade de São Paulo, São Paulo, SP, Brazil*
[c] *Unit of Rheumatology, Department of Paediatrics, Santa Casa de Misericórdia de São Paulo, São Paulo, SP, Brazil*
[d] *Unit of Paediatric Rheumatology, Universidade Estadual de Campinas, Campinas, SP, Brazil*
[e] *Service of Immunology, Allergy and Rheumatology, Department of Puericulture and Paediatrics, Faculdade de Medicina de Ribeirão Preto, Universidade de São Paulo, Ribeirão Preto, SP, Brazil*
[f] *Unit of Paediatric Rheumatology, Santa Casa de Misericórdia de Santos, Santos, SP, Brazil*
[g] *Service of Paediatric Rheumatology, Hospital Estadual de Bauru, Faculdade de Medicina de Botucatu, Botucatu, SP, Brazil*
[h] *Discipline of Paediatric Rheumatology, Faculdade de Medicina de Botucatu, Botucatu, SP, Brazil*
[i] *Private practice only*

ARTICLE INFO

Article history:
Received 17 September 2012
Accepted 17 February 2013

Keywords:
Paediatric rheumatology
Epidemiology
Registry

ABSTRACT

Introduction: Paediatric rheumatology (PR) is an emerging specialty, practised by a limited number of specialists. Currently, there is neither a record of the profile of rheumatology patients being treated in Brazil nor data on the training of qualified rheumatology professionals in the country.
Objective: To investigate the profile of PR specialists and services, as well as the characteristics of paediatric patients with rheumatic diseases, for estimating the current state of rheumatology in the state of São Paulo.
Patients and methods: In 2010, the scientific department of PR of the Paediatric Society of São Paulo administered a questionnaire that was answered by 24/31 accredited specialists in PR practising in state of São Paulo and by 8/21 institutions that provide PR care.
Results: Most (91%) of the surveyed professionals practise in public institutions. Private clinics (28.6%) and public institutions (37.5%) reported not having access to nailfold capillaroscopy, and 50% of the private clinics reported not having access to acupuncture. The average duration of professional practise in PR was 9.4 years, and 67% of the physicians had attended postgraduate programmes. Seven (87.5%) public institutions perform teaching activities, in which new paediatric rheumatologists are trained, and five (62.5%) offer post-

* *Corresponding author.*
E-mail: lucia@arrudacampos.com (L.M.A. Campos).

graduate programmes. Two-thirds of the surveyed specialists use immunosuppressants and biological agents classified as "restricted use" by the Health Secretariat. The disease most frequently reported was juvenile idiopathic arthritis (29.1-34.5%), followed by juvenile systemic lupus erythematosus (JSLE) (11.6-12.3%) and rheumatic fever (9.1-15.9%). The incidence of vasculitis (including Henoch-Schönlein purpura, Wegener's granulomatosis, and Takayasu's arteritis) and autoinflammatory syndromes was higher in public institutions compared to other institutions (P = 0.03, P = 0.04, P = 0.002, and P < 0.0001, respectively). Patients with JSLE had the highest mortality rate (68% of deaths), mainly due to infection.
Conclusion: The field of PR in the state of São Paulo has a significant number of specialists with postgraduate degrees who mostly practise at teaching institutions with infrastructures appropriate for the care of high-complexity patients.

Perfil de especialistas e de serviços em reumatologia pediátrica no estado de São Paulo

Palavras-chave:
Quadril
Adolescente
Criança
Condrólise idiopática de quadril

RESUMO

Introdução: A reumatologia pediátrica (RP) é uma especialidade emergente, com número restrito de especialistas, e ainda não conta com uma casuística brasileira sobre o perfil dos pacientes atendidos e as informações sobre a formação de profissionais capacitados.
Objetivo: Estudar o perfil dos especialistas e dos serviços em RP e as características dos pacientes com doenças reumáticas nessa faixa etária a fim de estimar a situação atual no estado de São Paulo (ESP).
Pacientes e métodos: No ano de 2010 o departamento científico de RP da Sociedade de Pediatria de São Paulo encaminhou um questionário respondido por 24/31 especialistas com título de especialização em RP que atuam no ESP e por 8/12 instituições com atendimento nesta especialidade.
Resultados: A maioria (91%) dos profissionais exerce suas atividades em instituições públicas. Clínicas privadas (28,6%) e instituições (37,5%) relataram não ter acesso ao exame de capilaroscopia e 50% das clínicas privadas não tem acesso à acupuntura. A média de tempo de prática profissional na especialidade foi de 9,4 anos, sendo 67% deles pós-graduados. Sete (87,5%) instituições públicas atuam na área de ensino, formando novos reumatologistas pediátricos. Cinco (62,5%) delas têm pós-graduação. Dois terços dos especialistas utilizam imunossupressores e agentes biológicos de uso restrito pela Secretaria da Saúde. A doença mais atendida foi artrite idiopática juvenil (29,1%-34,5%), seguida de lúpus eritematoso sistêmico juvenil (LESJ) (11,6%-12,3%) e febre reumática (9,1%-15,9%). Vasculites (púrpura de Henoch Schönlein, Wegener, Takayasu) e síndromes autoinflamatórias foram mais incidentes nas instituições públicas (P = 0,03; P = 0,04; P = 0,002 e P < 0,0001, respectivamente). O LESJ foi a doença com maior mortalidade (68% dos óbitos), principalmente por infecção.
Conclusão: A RP no ESP conta com um número expressivo de especialistas pós-graduados, que atuam especialmente em instituições de ensino, com infraestrutura adequada ao atendimento de pacientes de alta complexidade.

Introduction

Although paediatric rheumatology (PR) is an emerging specialty undergoing patent growth, the number of specialists in the field remains limited. Training of specialists and frequent revisions are needed to accommodate the expanding scope of rheumatic diseases and novel treatments.

The current state of this specialty, which is devoted to the knowledge of rheumatic diseases in childhood and adolescence, would benefit from a record of Brazilian rheumatology cases that would a establish a patient profile for this field. Additionally, little is known about the training of professionals qualified to assist children with rheumatic diseases in Brazil.

More than 20 years ago, the Paediatric Society of São Paulo (*Sociedade de Pediatria de São Paulo*, SPSP) established a department of PR, largely comprising university-based professionals with the task of providing updates, offering continuing education, and promoting a forum for the discussion of approaches, routines, and protocols to improve the care of children with rheumatic diseases.

To establish the current state of PR in the state of São Paulo, our aims were to use a survey to determine the profile of PR specialists and services, as well as establish the characteristics of paediatric patients exhibiting rheumatic diseases.

Patients and methods

In 2010, the management unit of the scientific department of PR of SPSP conducted a questionnaire for paediatricians who practise in the state of São Paulo and perform PR-related professional activities in private practices, clinics, outpatient facilities, and private or public institutions. The questionnaire was made available to the remainder of the scientific department members, who could then suggest changes before the final approval.

The questionnaire was sent electronically to 31 accredited PR professionals who practise in the state of São Paulo. The first part of the questionnaire comprised questions on the care that those specialists provide in the private workplaces: total number of patients cared for, number of patients per specific disease (including patients receiving follow-up care and new patients registered in 2010), occurrence of deaths in 2009 and 2010 (number of cases and cause of death), total duration of work in PR, weekly time devoted to PR, availability of support resources (hospitals for admissions, use of additional diagnostic tests, multiprofessional teams, and access to physicians practising other specialties), use of biological agents, use of invasive procedures, and participation in scientific meetings.

The second part of the questionnaire targeted only the individuals in charge of the institutions that provide PR care and included questions relative to the services provided: the type of institution (university-related, public or private and whether the institution performs teaching activities or not), length of time since establishment, total number of patients, number of patients per specific disease (including patients receiving follow-up care and new patients registered in 2010), occurrence of deaths in 2009 and 2010 (number of cases and cause of death), number of hours per week devoted to outpatient care, availability of support resources (number of beds for hospitalisation, use of additional diagnostic tests, multiprofessional teams, access to physicians practising other specialties, and presence of a research centre within the institution), use of biological agents, availability of diagnostic and therapeutic resources, use of invasive procedures, and participation in teaching activities (number of accredited PR specialists, residents, trainees, and current and previous postgraduate students trained at the institution).

The physicians were given two months to answer and return the questionnaires to the management unit of the scientific department of PR of SPSP (MTT, LMAC, EMO) for data analysis.

Results

The questionnaire was sent to 31 specialists in PR who practise in the state of São Paulo, seven of whom did not respond to the survey. Thus, 24 professionals (77.4%) participated in the present study. Data from eight public institutions in the state of São Paulo that provide PR care were collected (including six university-based services and two public hospitals, one of which performs teaching activities).

Among the professionals who participated in the study, 18 (75%) practised at university-based services and private clinics, two (8.3%) at university-based services only, two (8.3%) in private clinics only, one (4.2%) in a public hospital with teaching activities and in a private clinic, and one (4.2%) in a public hospital without teaching activities.

Among the 21 professionals who practise in private clinics, three (14.3%) reported attending both adult and paediatric patients with rheumatic diseases, and 18 (85.7%) reported attending paediatric patients only. Eleven (52.4%) professionals practised general paediatrics and paediatric rheumatology in their private practice. All of these professionals reported having access to hospitals to admit patients, as well as laboratories that perform clinical and rheumatologic tests and diagnostic imaging facilities (simple radiographs, ultrasonography, computed tomography, nuclear magnetic resonance, and bone densitometry). Six (28.6%) paediatric rheumatologists reported no access to nailfold capillaroscopy. Twelve (57.1%) of the professionals indicated performing joint injections.

Eighteen (85.7%) out of the 21 professionals who practised in private clinics reported performing intravenous infusions of corticosteroids and immunosuppressants, and 14 (66.7%) prescribe biological agents. Seven (50%) out of the 14 paediatric rheumatologists who use biological agents perform the infusions in the day-hospital setting. The available multiprofessional teams included physical therapists (95.8%), nutritionists (87.5%), psychologists (83.3%), speech therapists (66.7%), and dentists (50%). Other available medical specialties included neurology (100%), ophthalmology (100%), nephrology (100%), orthopaedics (100%), paediatric surgery (95.8%), cardiology (91.7%), dermatology (91.7%), haematology (91.7%), vascular care (79.2%), intensive care (79.2%), and acupuncture (50%).

The duration of PR practise of the physicians in the public institutions and private clinics varied from 0.5 to 26 years (9.4 years on average). Among the 24 specialists, three were full professors, two were professors, eight held PhD degrees, three held MA degrees, and eight had not attended a stricto sensu postgraduate programme. All the professionals but one (95.8%) reported attending national or international scientific meetings for continuing education in the past two years.

All the investigated institutions have outpatient facilities and inpatient beds. The outpatient workload varies from four to 24 hours per week. Three (37.5%) of the participating institutions have their own rheumatology laboratory, and three (37.5%) do not have the means to perform nailfold capillaroscopy. Five institutions (62.5%) perform joint injections, and three (37.5%) perform biopsies. Seven (87.5%) out of the eight surveyed institutions use biological agents with their patients. All eight institutions have multiprofessional teams and provide access to other medical specialists for advice.

Seven (87.5%) public institutions perform teaching activities, including training of new paediatric rheumatologists, general paediatric residents and trainees, and PR residents and physicians seeking accreditation. Five (62.5%) out of the

Table 1 – Characteristics of the teaching activities relative to paediatric rheumatology in public institutions in 2010 and since the establishment of the institutions.

Characteristics	2010 (n)	Total[a] (n)
Institutions with teaching activities	7	7
Institutions with postgraduate programmes	5	5
PR accredited specialists	29	48
PR residents	9	50
PR trainees	10	118
Postgraduate PR students	27	72
Paediatric rheumatologists with simultaneous private practices	18	18

PR, paediatric rheumatology.
[a]Total number since the establishment of the institutions 6.

eight surveyed institutions offer postgraduate programmes (Table 1). Six (75%) institutions have a general research centre, and three (37.5%) have a research centre specialising in rheumatology.

The PR services at the public institutions have been in operation for 22 years on average (varying from one to 43 years), and the total number of patients with rheumatic diseases diagnosed and treated since the establishment of these institutions was 19,078. The PR services at the private institutions have been in operation for 7.3 years on average (varying from 0.5 to 25 years), and the total number of patients with rheumatic diseases diagnosed and treated since the establishment of these institutions was 5,094. Thus, the total number of patients seen at both public institutions and private clinics was 24,172. In 2010, which was the last year considered in the present study, 1,427 new cases were diagnosed (Table 2). The incidence of juvenile idiopathic arthritis and rheumatic fever was greater in private clinics, while the incidence of Henoch-Schönlein purpura, Wegener's granulomatosis, Takayasu's arteritis, and autoinflammatory syndromes was greater in public institutions.

Nine deaths occurred in 2009 (all corresponding to patients cared for at public institutions), and 13 occurred in 2010 (12 were patients cared for at public institutions) (Table 3).

Table 2 – New cases of rheumatic disease in public institutions and private clinics in 2010.

Disease	Public institutions (n = 519)	Private clinics (n = 908)	Total (n = 1427)	P
Juvenile idiopathic arthritis	151 (29.1)	313 (34.5)	464	0.037
Juvenile systemic lupus erythematosus	60 (11.6)	112 (12.3)	172	0.665
Rheumatic fever	47 (9.1)	144 (15.9)	191	0.0003
Juvenile dermatomyositis	25 (4.8)	33 (3.6)	58	0.276
Henoch-Schönlein purpura	145 (27.9)	207 (22.8)	352	0.030
Kawasaki disease	17 (3.3)	37 (4.0)	54	0.446
Polyarteritis nodosa	9 (1.7)	7 (0.8)	16	0.096
Wegener's granulomatosis	4 (0.8)	1 (0.1)	5	0.042
Takayasu's arteritis	9 (1.7)	2 (0.2)	11	0.002
Juvenile scleroderma	24 (4.6)	41 (4.5)	65	0.924
Autoinflammatory syndromes	28 (5.4)	11 (1.2)	39	< 0.0001

Data expressed as n (%).

Table 3 – Cause of death in patients with rheumatic disease in 2009 and 2010.

Cause	2009 (n = 9)	2010 (n = 13)
Infection	4 JSLE/1 JDM/ 1 JIA	4 JSLE
Disease activity	1 JDM/1 JIA	4 JSLE/1 JIA/ 1 TA/1 RF
Disease activity and infection	1 JSLE	2 JSLE

JSLE, juvenile systemic lupus erythematosus; JDM, juvenile dermatomyositis; JIA, juvenile idiopathic arthritis; TA, Takayasu's arteritis; RF, rheumatic fever.

Discussion

The aim of the present study was to assess the qualification of practising PR professionals, the scope of care provided, and the specific PR diagnoses in the state of São Paulo. Although seven out of 31 professionals did not answer the questionnaire, the population sample is quite representative of the PR specialists currently practising in São Paulo, as the responders comprised more than three-fourths of the original population who had received the questionnaire.

The completed questionnaires revealed that most PR professionals practise in public institutions, either exclusively or in conjunction with another type of institution. This finding reflects the significant complexity that characterises PR, which demands an interface with other healthcare professionals and the promotion of continuing education from its specialists.

Because all the participants included in the study are paediatricians, most of the professionals who responded to the questionnaire see children and adolescents exclusively. The fact that half of the participants do not see patients with rheumatic diseases exclusively in their private practices might be due to several reasons: the number of patients with rheumatic diseases represents a small fraction of the population that seeks paediatric care, and the high cost of treatment and follow-up for this type of patient frequently compels physicians to refer such patients to public institutions. A large fraction of the patients require referral to tertiary centres, due to the need for advanced diagnostic methods and complex treatments. The majority of such institutions afford the human and material resources needed for PR care. A survey conducted in North America showed that three-fourths of paediatric rheumatologists devote at least 90% of their time to the care of children and adolescents.[1]

Limited access to nailfold capillaroscopy compared to other diagnostic tests in both private clinics and public institutions is worth noting. Nailfold capillaroscopy has prov-

en useful in the diagnosis and follow-up of patients with rheumatic diseases in recent years, particularly in cases of systemic scleroderma and juvenile dermatomyositis.[2,3] The limited availability of this procedure in the state of São Paulo suggests a lack of equipment and training of specialists in this field, thus suggesting an available sub-speciality for the training of new professionals. Similarly, the PR professionals at private clinics have less access to services requiring greater specialisation and technical resources, such as acupuncture and intensive care.

With regard to treatment, approximately two-thirds of the surveyed specialists use drugs classified as "restricted use" (due to their high cost) by the Health Secretariat, such as immunosuppressants and biological agents, which have proven to be highly beneficial in recent years for the control of disease activity in rheumatic patients.[4-6] One of the latest therapeutic innovations for such patients is the intravenous administration of such drugs in the day-hospital setting, which was reported to be used by 50% of the professionals who participated in the present study, as this procedure shortens the duration of the hospital, with a consequent reduction in the risk of infection.

Although the surveyed professionals practised PR for an average of 9.4 years, all but one participated in scientific meetings for the purpose of continuing their education in the past two years; this finding corroborates the need for continuing education in this field on a steady basis, even in the case of the professional with significant experience and high academic degrees. With regard to the training of new professionals, almost all the surveyed public institutions perform teaching and research activities, and a significant number of students were enrolled in postgraduate programmes in PR in the state of São Paulo, with many of these students coming from other states.

Although there are no data on the incidence and prevalence of rheumatic diseases in Brazil as a whole, or in the state of São Paulo in particular, juvenile idiopathic arthritis (JIA) was the disease most frequently reported by the surveyed paediatric rheumatologists, followed by juvenile systemic lupus erythematosus (JSLE), in accordance with the international literature.[7-9] Notably, although rheumatic fever (RF) was commonly the most prevalent rheumatic disease in developing countries in the past,[10] the frequency of this disease has been decreasing in the state of São Paulo. This observation may reflect the overall improvement in the population's living conditions and greater awareness by healthcare professionals in the prevention of RF. Recently, the Health Ministry published guidelines for the diagnosis and treatment of RF, representing one additional tool for the prevention of this disease by primary prophylaxis and for more accurate diagnosis and treatment of RF by professionals in the primary healthcare network.[11] Surprisingly, the incidence of RF was greater in private clinics compared to public institutions; in contrast, the incidence of vasculitis and autoinflammatory syndromes was greater in the latter, probably as a function of the complexity of the reported cases.

In the present survey, JSLE was associated with the highest mortality rate, corresponding to 68% of the deaths of patients with rheumatic diseases in 2009 and 2010. The medical literature corroborates the severity of this condition and its high mortality risk.[12,13] The main cause of death of those patients with JSLE was infection, either alone or associated with disease activity, in accordance with the literature.[14] Most deaths occurred at public institutions, which may be attributable to delayed diagnosis and consequent poorer prognosis and referral to tertiary centres and to the greater referral of more severe cases to this type of institution.

The present study had some limitations, such as the inability to survey all PR professionals in the state of São Paulo; the questionnaire was sent only to the professionals registered as specialists at SPSP, thus excluding adult rheumatologists who also see children and adolescents. It was also not possible to assess the complete population of paediatric patients with rheumatic diseases, as a fraction of these patients were cared for at primary healthcare services or by other types of specialists, such as orthopaedists, general paediatricians, and adult rheumatologists. In addition, there were limitations inherent to the methods used (i.e., questionnaires sent by e-mail), despite prior discussions and presentations of the protocol at meetings held by the PR scientific department of SPSP and the direct contact with the professionals, whose answers raised doubts at the time of tabulating the data. Finally, patients who may have visited two different physicians or institutions might represent a source of bias in this study due to the involuntary duplication of the patients' information, as the data were collected anonymously and the study had a retrospective design.

We conclude that the field of PR in the state of São Paulo includes a significant number of specialists and postgraduate professionals, who mainly practise at teaching institutions with infrastructures appropriate for the care of high-complexity patients. The shift in the prevalence of rheumatic diseases in the state of São Paulo reflects the changes in this population's living conditions, as well as in healthcare services. With the progressive increase in the occurrence of chronic diseases in the paediatric population, services specialised in the various corresponding fields are needed to allow for a timely referral of the affected patients.

This pilot study was designed with the expectation that it might be extended to the national level to establish the current state of Brazilian PR, identify geographical areas that lack sufficient numbers of professionals in this field, and identify the needs and conditions of each service currently being offered for the care of rheumatic patients. Such findings would provide objective data for planning a more adequate allocation of human and material resources to meet the demands of rheumatic disease treatment.

REFERENCES

1. Henrickson M. Policy challenges for the pediatric rheumatology workforce: Part II. Health care system delivery and workforce supply. Henrickson Pediatric Rheumatology 2011;9(1):24.
2. Nascif AK, Terreri MT, Len CA, Andrade LE, Hilário MO. Inflammatory myopathies in childhood: correlation between nailfold capillaroscopy findings and clinical and laboratory data. J Pediatr. (Rio J). 2006;82(1):40-5.
3. Ingegnoli F, Zeni S, Gerloni V, Fantini F. Capillaroscopic observations in childhood rheumatic diseases and healthy controls. Clin Exp Rheumatol. 2005;23(6):905-11.

4. Hayward K, Wallace CA. Recent developments in anti-rheumatic drugs in pediatrics: treatment of juvenile idiopathic arthritis. Arthritis Res Ther. 2009;11(1):216.
5. Gurion R, Lehman TJ, Moorthy LN. Systemic arthritis in children: a review of clinical presentation and treatment. Int J Inflam. 2012; 271569. [Published online 2011 December 25] doi:10.1155/2012/271569.
6. Woo P. Theoretical and practical basis for early aggressive therapy in pediatric autoimmune disorders. Curr Opin Rheumatol. 2009;21(5):552-7.
7. Huemer C, Huemer M, Dorner T, Falger J, Schacherl H, Bernecker M, et al. Incidence of pediatric rheumatic diseases in a regional population in Austria. J Rheumatol. 2001;28(9):2116-9.
8. Denardo BA, Tucker LB, Miller LC, Szer IS, Schaller JG. Demography of a regional pediatric rheumatology patient population. Affiliated Children's Arthritis Centers of New England. J Rheumatol. 1994;21(8):1553-61.
9. Rosenberg AM. Longitudinal analysis of a pediatric rheumatology clinic population. J Rheumatol. 2005;32(10):1992-2001.
10. Sztajnbok FR, Serra CR, Rodrigues MC, Mendoza E. Rheumatic diseases in adolescence. J Pediatr. (Rio J) 2001;77(Supl.2):s234-s44.
11. Barbosa PJ, Muller RE, Latado AL, Achutti AC, Ramos AIO, Weksler C, et al. Diretrizes brasileiras para diagnóstico, tratamento e prevenção da febre reumática da Sociedade Brasileira de Cardiologia, da Sociedade Brasileira de Pediatria e da Sociedade Brasileira de Reumatologia. Arq Bras Cardiol. 2009;93(3 supl.4):1-18.
12. Hashkes PJ, Wright BM, Lauer MS, Worley SE, Tang AS, Roettcher PA, et al. Mortality outcomes in pediatric rheumatology in the US. Arthritis Rheum. 2010; 62(2):599-608.
13. Tucker LB, Uribe AG, Fernández M, Vilá LM, McGwin G, Apte M, et al. Adolescent onset of lupus results in more aggressive disease and worse outcomes: results of a nested matched case-control study within LUMINA, a multiethnic US cohort (LUMINA LVII). Lupus. 2008;17(4):314-22.
14. Faco MM, Leone C, Campos LM, Febrônio MV, Marques HH, Silva CA. Risk factors associated with the death of patients hospitalized for juvenile systemic lupus erythematosus. Braz J Med Biol Res. 2007;40(7):993-1002.