EDITORIAL

Editorial

Is cervical spine involvement in juvenile polyarthritis under-recognized?

Detecting cervical abnormalities early

This editorial refers to Radiological cervical spine involvement in young adults with polyarticular juvenile idiopathic arthritis, by M Elhai, J Wipff, R Bazeli *et al.*, doi:10.1093/rheumatology/kes054, on pages 267-275.

Elhai *et al.*'s [1] study evaluates the frequency of radiological cervical spine involvement in young adults with polyarticular JIA (pJIA) compared with RA patients. The authors show that about two-thirds of patients in both groups had radiographic abnormalities in the cervical spine, including anterior atlantoaxial subluxation, erosion of the odontoid process, C1–C2 arthritis and apophyseal joint arthritis. Lesions were found even in patients without clinical symptoms and their presence correlated positively with the severity of disease.

The similar prevalence of structural cervical spine lesions in young adults with pJIA and RA highlights the importance of this complication for the long-term follow-up of juvenile polyarthritis. This aspect of pJIA outcome needs further study because literature on the topic is relatively scarce, in contrast to the abundant number of papers on cervical problems in RA. Another striking point of this study is the high percentage of asymptomatic patients presenting with radiological cervical lesions. Only 42% of patients without neck complaints or findings on examination had a normal cervical scan; accordingly, the authors raise the question of the frequency of systematic cervical spine radiological controls during follow-up in pJIA.

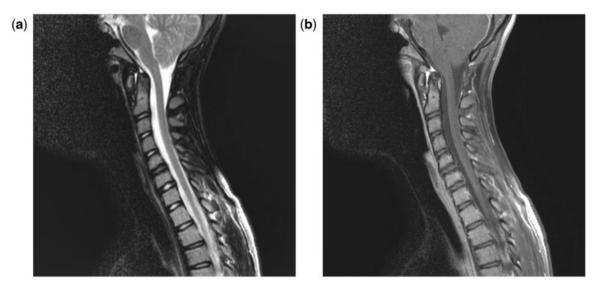
According to the ILAR classification [2], polyarthritis patients with onset before the age of 16 years are divided into two different categories, depending on the result of the RF [2]. RF-negative polyarthritis corresponds to a heterogeneous group of patients with symmetrical or asymmetrical joint involvement, presence or absence of ANAs, and includes 10-12% of the JIA cases [3, 4]. The RF-positive polyarticular category is considered to be an early form of RA and represents 2-3% of JIA patients [3]. The long-term prognosis of pJIA patients can vary but outcome studies with a follow-up of at least 10 years have shown that active arthritis is still present in 39-67% of patients [5]. In particular, the latter study showed that children who did not present with a period of disease inactivity in the 5 years of disease onset had more long-term joint damage. In another study, patients with RF-positive polyarthritis or early evidence of joint damage had a higher risk of persistent active arthritis during follow-up [6].

In Elhai et al.'s [1] study, RF-positive patients represented 60% of the cohort: more than 10 times the expected frequency in normal JIA distribution, indicating an over-representation of this category. The high proportion of RF-positive patients is likely to have influenced the results of the study and could explain similar cervical spine involvement in JIA and RA patients. Interestingly, the frequency of a positive RF was similar in patients with or without cervical involvement; these data suggest that RF-negative patients had a similarly high rate of cervical spine abnormalities, which may be explained by a significantly longer duration of disease course in comparison with that in RF-positive patients (17.3 vs 9.8 years).

Imaging evaluation of children with polyarthritis is usually indicated according to clinical findings, mainly in patients with joint inflammation that persists through treatment. Plain radiography is the standard imaging technique for monitoring abnormalities in peripheral joints, but there are no recommendations for timing intervals during the follow-up of JIA patients [7]. Cervical spine involvement has been described in different forms of JIA, including polyarthritis, systemic arthritis and enthesitis-related arthritis. In 2002, Laiho et al. [8] reported cervical spine structural abnormalities in 62% of 159 JIA patients, who underwent radiographic examination because of neck symptoms or as a part of a pre-operative routine. More than 80% of the patients suffered from polyarthritis, and ~25% of them were RF positive. Elhai et al.'s [1] study had similar findings but the patient cohort was guite different, since two-thirds of the patients were treated with a biotherapy and half of them did not show any neck symptoms or signs.

Many children and adolescents with rheumatic diseases are treated by paediatricians who are specialists in rheumatology in paediatric settings; care for young patients will be adjusted to factors related to their age, like growth, family, and school. The paediatrician will transfer the patient to an adult clinic at the end of their adolescence and will not see their long-term outcome. Transition clinics have recently been developed to offer a smooth transfer to adult care, which is essential for continuity of care. Transition clinics have another major advantage in that they highlight the outcome of JIA patients during early adulthood. The information gathered from young adult JIA patients is useful in improving the care provided to children with JIA. Elhai et al.'s [1] study shows that cervical involvement in pJIA is common and can occur even in the absence of symptoms. This finding underlines the importance for clinicians of detecting minor cervical symptoms in JIA patients. The bias of the study could be that subjects with mild disease, which can go into long-term

Fig. 1 MRI with cervical involvement.



(a) Without contrast (T1-weighted) and (b) with contrast (T2 fat-saturation) of a 15-year-old girl with pJIA: periodontoid effusion, vertebral bone abnormalities and articular contrast enhancement.

remission, are less likely to seek medical care. In light of this, should pJIA patients with only minor cervical symptoms still undergo regular cervical imaging? Prospective trials need to be done before we can make clear recommendations.

In Elhai et al.'s [1] study, mean disease duration before the introduction of the biological agents was 8.2 years, since biotherapies were not licensed for JIA at disease onset in the majority of the patients studied. These new treatments have improved the control of joint inflammation and we expect that they will decrease the high incidence of cervical spine involvement in future. Finally, even if plain X-rays are easy to obtain, newer imaging techniques, such as MRI with contrast, could be useful as they are more sensitive in detecting joint inflammation (Fig. 1) and pre-destructive changes [7]. They are now the method of choice in detecting cervical spine involvement in patients with pJIA. Early diagnosis of cervical abnormalities would be important in selecting pJIA patients to be more aggressively treated and for whom protective measures should be implemented.

Disclosure statement: The authors have declared no conflicts of interest.

Michaël F. Hofer^{1,2} and Rolando Cimaz³

¹Paediatric Rheumatology of Western Switzerland, Paediatric Department, CHUV, University Hospital, Lausanne,
²Paediatric Department, HUG, Geneva, Switzerland and
³AOU Meyer, Firenze, Italy.
Accepted 6 September 2012
Correspondence to: Michaël F. Hofer, Paediatric
Rheumatology, Department of Paediatrics, BH11-CHUV,
CH-1011 Lausanne, Switzerland.
E-mail: michael.hofer@chuv.ch

References

- Elhai M, Wipff J, Bazeli R et al. Radiological cervical spine involvement in young adults with polyarticular juvenile idiopathic arthritis. Rheumatology 2012, Advance Access published 17 April 2012 doi:10.1093/ rheumatology/kes054.
- 2 Petty RE, Southwood TR. International League of Associations for Rheumatology classification of juvenile idiopathic arthritis: second revision, Edmonton, 2001. Rheumatology 2004;31:2.
- 3 Modesto C, Antón J, Rodriguez B et al. Incidence and prevalence of juvenile idiopathic arthritis in Catalonia (Spain). Scand J Rheumatol 2010;39:472–9.
- 4 Ravelli A, Varnier GC, Oliveira S *et al*. Antinuclear antibody-positive patients should be grouped as a separate category in the classification of juvenile idiopathic arthritis. Arthritis Rheum 2011;63:267-75.
- 5 Magnani A, Pistorio A, Magni-Manzoni S *et al*. Achievement of a state of inactive disease at least once in the first 5 years predicts better outcome of patients with polyarticular juvenile idiopathic arthritis. J Rheumatol 2009;36:628–34.
- 6 Ringold S, Kristy D, Seidel K, Koepsell T, Wallace CA. Inactive disease in polyarticular juvenile idiopathic arthritis: current patterns and associations. Rheumatology 2009; 48:972–7.
- 7 Breton S, Jousse-Joulin S, Finel E *et al.* Imaging approaches for evaluating peripheral joint abnormalities in juvenile idiopathic arthritis. Semin Arthritis Rheum 2012; 41:698-711.
- 8 Laiho K, Savolainen A, Kautiainen H, Kekki P, Kauppi M. The cervical spine in juvenile chronic arthritis. Spine J 2002;2:89-94.