

RESEARCH

Open Access



# Development of a scoring system to assist clinicians in the early referral of patients with suspected juvenile idiopathic arthritis: the EasyJIA score

Achille Marino<sup>1\*</sup>, Carlo Alberto Scirè<sup>2,3</sup>, Paola Baldassarre<sup>4</sup>, Cristina Ferrigno<sup>4</sup>, Stefania Costi<sup>1</sup>, Francesco Baldo<sup>1</sup>, Maurizio Virgilio Gattinara<sup>1</sup>, Davide Rozza<sup>5</sup>, Cecilia Beatrice Chighizola<sup>1,6</sup> and Roberto Felice Caporali<sup>1,6</sup>

## Abstract

**Objective** Juvenile idiopathic arthritis (JIA) is the most common chronic pediatric rheumatic disease. Early referral to a specialized center is crucial for prompt diagnosis and treatment. This study aims to develop and validate a scoring system to assist clinicians in efficiently identifying and referring patients suspected of having non-systemic JIA.

**Methods** We conducted a cohort study with a mixed design (retrospective and prospective), involving consecutive patients presenting with joint complaints who were referred for the first time to the Pediatric Rheumatology Unit at ASST G. Pini-CTO Hospital. The model was developed using multivariate logistic regression with bootstrap resampling and the Lasso (Least Absolute Shrinkage and Selection Operator) method for variable selection.

**Results** A total of 342 patients were included, of whom 61 (18%) were diagnosed with JIA. The selected variables for the model were: type of joint (large), daily symptoms, joint swelling, activity as a precipitating factor, a positive squeeze test of the metatarsophalangeal/metacarpophalangeal (MTP/MCP) joints, normal bending of the interphalangeal (IF) joints of the hands, morning limping and/or stiffness, and sacroiliac tenderness. The ROC curve, based on the model's regression score, showed an AUC of 0.92 with an overall accuracy of 0.88 (95% CI: 0.84–0.91) using a cutoff of 3 points, yielding a sensitivity of 95% and a specificity of 71%. Initial internal validation of the model revealed an AUC of 0.92 (95% CI: 0.89–0.95).

**Conclusion** This study presents and initially validates a simple and efficient scoring system to aid clinicians in the early referral of patients suspected of having non-systemic JIA.

**Clinical trial number** Not applicable.

\*Correspondence:

Achille Marino  
achille.marino@asst-pini-cto.it

<sup>1</sup>Unit of Pediatric Rheumatology, ASST G. Pini-CTO, Piazza Cardinal Ferrari 1, Milan 20122, Italy

<sup>2</sup>Rheumatology Unit, Fondazione IRCCS San Gerardo dei Tintori, Monza, Italy

<sup>3</sup>School of Medicine, University of Milano-Bicocca, Milan, Italy

<sup>4</sup>Department of Biomedical and Clinical Sciences, Buzzi Children's Hospital, University of Milan, Milan, Italy

<sup>5</sup>Epidemiology Unit, Italian Society for Rheumatology (SIR), Milan, Italy

<sup>6</sup>Department of Clinical Sciences and Community Health, Research Center for Pediatric and Adult Rheumatic Diseases (RECAPRD), University of Milan, Milan, Italy



© The Author(s) 2026. **Open Access** This article is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License, which permits any non-commercial use, sharing, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if you modified the licensed material. You do not have permission under this licence to share adapted material derived from this article or parts of it. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit <http://creativecommons.org/licenses/by-nc-nd/4.0/>.

## Introduction

Juvenile idiopathic arthritis (JIA) is the most frequent chronic pediatric rheumatic disease, with a prevalence varying between 16 and 150 per 100,000 with a pooled annual incidence of 7.8 per 100,000 population [1, 2]. JIA is not a single entity but refers to arthritis, lasting for at least 6 weeks, without an identified etiology, with onset before age 16 [3]. The term JIA encompasses a heterogeneous group of diseases currently classified into clinical categories according to the International League of Associations for Rheumatology (ILAR) classification system: oligoarticular JIA, rheumatoid factor (RF) negative polyarticular JIA, RF positive polyarticular JIA, psoriatic arthritis, enthesitis-related arthritis (ERA), systemic JIA, and undifferentiated JIA [3]. JIA categories vary in terms of disease severity, clinical manifestations, response to treatment, and long-term consequences [3, 4]. Clinical presentation is usually dominated by joint swelling and limitation, which are typically more relevant than pain.

Currently, in the absence of pathognomonic features for JIA, the diagnosis relies on specialist expertise and the exclusion of other conditions.

The diagnosis of JIA requires thorough medical history and a comprehensive physical examination, including a detailed joint assessment. The concept of a “window of opportunity” to modify the natural course of JIA and improve patient outcomes has become increasingly accepted, making early referral to pediatric rheumatology (PR) centers and the timely initiation of appropriate therapy critical [5, 6]. However, studies worldwide indicate that, despite variations in healthcare access, children with JIA often experience significant delays in being referred to PR centers, mainly due to prolonged referral processes following the initial contact with primary care providers [7–9].

Timely referral to a specialized center is essential for early diagnosis, altering the disease’s progression, improving quality of life, and reducing the risk of joint damage and disability [5, 6, 10, 11]. Delays and uncertainties in diagnosis can also have a negative psychological impact on both JIA patients and their families [10–12]. In many countries, healthcare professionals express discomfort with performing rheumatologic assessments and addressing musculoskeletal complaints [13]. Therefore, establishing fast-track pathways for timely referral to specialized care settings is imperative. This study aims to develop and validate a scoring system based on medical history and simple clinical assessments to help clinicians efficiently identify and refer patients suspected of having non-systemic JIA to pediatric rheumatology specialists.

## Methods

This cohort study included consecutive patients presenting with joint complaints who were referred for the first time to the Pediatric Rheumatology Unit at ASST G.Pini-CTO Hospital between April 1, 2022, and July 31, 2024. This is a cohort study with mixed design: data up to May 31, 2023, were collected retrospectively, with prospective data collection occurring thereafter.

Inclusion criteria: joint complaints as reasons of referral, under 16 years old at the time of referral. Joint complaints were defined as the presence of joint pain, swelling, stiffness, or any combination of these symptoms. Exclusion criteria: presence of fever at the time of referral, pre-existing conditions that may affect musculoskeletal functions and examination.

During the initial evaluation at our center, we gathered the patient’s medical history through a customized questionnaire (supplemental material), focusing on the frequency and patterns of joint complaints, triggers, presence of back pain and related symptoms, morning limping or stiffness, visible joint swelling and its patterns, the capability of performing simple maneuvers (e.g. “making a fist”), and any accompanying constitutional symptoms. All recorded data were collected during routine visits, and patients were monitored until their final diagnosis was reached. Patients were diagnosed with JIA according to the ILAR classification criteria [3]. Compliance with the Health Insurance Portability and Accountability Act of 1996 and the principles of the Declaration of Helsinki was maintained throughout the study. Approval was granted by our center’s Institutional Review Board (IRB) (4085\_S\_P). Informed consent for medical chart data collection was obtained from all patients or their legal guardians.

Quantitative variables were summarized as medians with range, or interquartile range (IQR), while categorical variables were expressed as absolute and relative frequencies. The Shapiro-Wilk test was used to assess the normality of distributions. Differences in quantitative variables were analyzed using the non-parametric Mann-Whitney U test. Associations between categorical variables were evaluated with either the Chi-Square test or Fisher’s exact test, as appropriate.

Due to the presence of some missing values, which were assumed to be missing completely at random (MCAR) or missing at random (MAR), we applied multiple imputations using a chained equations approach to make the most efficient use of the available data. We generated five imputed datasets with a maximum of 100 iterations, using the mice package (RStudio). Quantitative variables were imputed via predictive mean matching (pmm), while categorical variables were assigned using Classification and Regression Trees (CART), which do not require underlying assumptions.

The variable selection process for the multivariate logistic analysis involved three steps:

- Bootstrap Resampling: we initially applied the bootstrap method to evaluate the variability and robustness of the variables. This involved repeatedly resampling the data and assessing the model's performance to determine which variables consistently contributed to the model.
- Lasso Regression: next, we used Lasso (Least Absolute Shrinkage and Selection Operator) for variable selection. This technique creates a subset of important variables and mitigates overfitting by incorporating a penalty term that drives some coefficients to zero, effectively performing variable selection and regularization. The overall process involves running LASSO multiple times on bootstrapped samples and then calculating the frequency with which each variable is selected.
- Clinical Judgment: to enhance the positive predictive power, we selected variables for inclusion in the multivariate analysis based on clinical judgment. This selection was made from variables with a selection frequency exceeding 250 (or 50%) in the bootstrap and Lasso analysis.

This approach ensures a robust and clinically relevant selection of variables for the logistic regression model. To assess its discriminative ability, the receiver operating characteristic (ROC) curve and the area under the curve (AUC) were derived from the model.

To create a simplified score for routine clinical use, we rounded the regression coefficients from the final multivariable logistic regression model to the nearest integer. The ROC curve of this simplified model was then compared with that of the original model to evaluate any differences in predictive accuracy. To further validate the model, we applied the bootstrap approach once more, reinforcing the reliability of the results through resampling techniques.

A significance threshold of  $P \leq 0.05$  was applied. All analyses were performed using RStudio (version 2021.09.2 + 382 for macOS).

## Results

### General features of the cohort

In this study, we included a total of 342 patients with joint complaints who were referred to our center, of whom 61 (18%) were diagnosed with JIA. A diagnosis of non-inflammatory joint pain was established in 229 (67%) subjects of the overall cohort, which included 26 (11.5%) individuals with amplified musculoskeletal pain syndrome. Inflammatory conditions other than JIA were diagnosed in 15% of cases, with reactive arthritis being

the most common (20 patients, 38%). Among the JIA patients, the most common subtype was oligoarticular (38 patients, 62%), followed by polyarticular (15 patients, 25%), and enthesitis-related arthritis (8 patients, 13%). The demographic and key clinical characteristics of the cohort are summarized in Table 1.

There were no significant differences in age at the initial PR evaluation, sex distribution, or prevalence of arthralgia between JIA and non-JIA patients, time lag between symptom onset and the first healthcare consultation (Table 1). Similarly, the localization of symptoms was comparable between the two groups (Table 1). However, monoarticular involvement tended to be more frequently among JIA patients compared to non-JIA individuals (39% vs. 27%,  $p = 0.06$ ).

In contrast, the daily patterns of joint complaints differed significantly between the groups. JIA patients experienced daily symptoms and morning stiffness and/or limping more frequently (74% vs. 27%;  $p < 0.01$ ). Interestingly, the duration of morning stiffness (MS) was directly associated with a higher likelihood of JIA, increasing from 13% among those with 15–30 min of MS to 25% in patients with over 1 h of MS. Night awakening due to joint pain was uncommon in both groups (15% vs. 19%;  $p = 0.5$ ).

Additionally, JIA and non-JIA patients exhibited opposing responses to physical activity and rest: JIA patients typically experienced symptom worsening after periods of rest, whereas non-JIA individuals were more likely to develop symptoms following physical activity (Table 1).

### Model development

Using the bootstrap method and LASSO regression, we generated a list of variables ranked by their frequency (detailed frequencies of the selected variables are provided in Table 4 of the supplemental material).

The process involves repeatedly running LASSO on bootstrapped samples and calculating how often each variable is selected. From the covariates with a frequency exceeding 250 (indicating choice in more than 50% of cases), we selected the following based on clinical judgment: joint type (large), daily symptoms, joint swelling, activity as a precipitating factor, a positive squeeze test of the metatarsophalangeal/metacarpophalangeal (MTP/MCP) joints, normal bending of the interphalangeal (IF) joints of the hands, morning limping and/or stiffness, and the presence of sacroiliac tenderness. The clinical items of the model are shown in Fig. 1. The results of the logistic regression model are presented in Table 2. The prediction model derived from the development data showed excellent discrimination, achieving an AUC of 0.92 (Fig. 2A). The Hosmer-Lemeshow test yielded a  $p$ -value of 0.88.

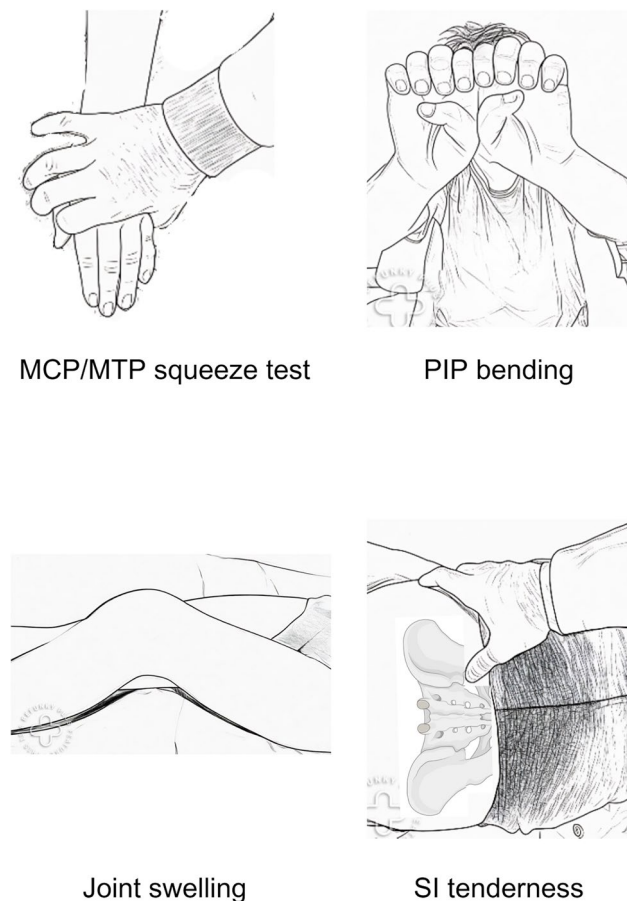
**Table 1** Key features of the cohort

|                                                                            | Overall<br>N=342 <sup>1</sup> | No JIA<br>N=281 <sup>1</sup> | JIA<br>N=61 <sup>1</sup> | p-value <sup>2</sup> |
|----------------------------------------------------------------------------|-------------------------------|------------------------------|--------------------------|----------------------|
| Age at first Pediatric Rheumatology evaluation, years                      |                               |                              |                          | 0.6                  |
| Median (IQR)                                                               | 10.1 (7.1–12.8)               | 10.0 (7.2–12.7)              | 10.8 (4.9–14.2)          |                      |
| Unknown                                                                    | 1                             | 1                            | 0                        |                      |
| Sex                                                                        |                               |                              |                          | 0.3                  |
| Female                                                                     | 194 (57%)                     | 156 (56%)                    | 38 (62%)                 |                      |
| Time lag between symptom onset and the first healthcare consultation, days |                               |                              |                          | 0.7                  |
|                                                                            | 14 (0–122.2)                  | 14 (0–182)                   | 14 (5–31.5)              |                      |
| Arthralgia                                                                 |                               |                              |                          | 0.2                  |
|                                                                            | 284 (83%)                     | 237 (84%)                    | 47 (77%)                 |                      |
| Joint swelling                                                             |                               |                              |                          | <0.01                |
|                                                                            | 119 (35%)                     | 72 (26%)                     | 47 (77%)                 |                      |
| Limping                                                                    |                               |                              |                          | 0.10                 |
|                                                                            | 79 (23%)                      | 60 (21%)                     | 19 (31%)                 |                      |
| Monoarticular involvement                                                  |                               |                              |                          | 0.06                 |
| Unknown                                                                    | 10                            | 8                            | 2                        |                      |
| Back involvement                                                           |                               |                              |                          | 0.5                  |
| Unknown                                                                    | 1                             | 0                            | 1                        |                      |
| Lower limb involvement                                                     |                               |                              |                          | 0.09                 |
|                                                                            | 292 (88%)                     | 244 (89%)                    | 48 (81%)                 |                      |
| Unknown                                                                    | 10                            | 8                            | 2                        |                      |
| Upper limb involvement                                                     |                               |                              |                          | 0.3                  |
|                                                                            | 132 (40%)                     | 105 (38%)                    | 27 (46%)                 |                      |
| Unknown                                                                    | 10                            | 8                            | 2                        |                      |
| Daily symptoms                                                             |                               |                              |                          | <0.01                |
|                                                                            | 146 (52%)                     | 95 (43%)                     | 51 (88%)                 |                      |
| Unknown                                                                    | 61                            | 58                           | 3                        |                      |
| Morning limping and/or stiffness                                           |                               |                              |                          | <0.01                |
|                                                                            | 122 (36%)                     | 77 (27%)                     | 45 (74%)                 |                      |
| Rest as a precipitating factor                                             |                               |                              |                          | <0.01                |
|                                                                            | 75 (25%)                      | 44 (18%)                     | 31 (54%)                 |                      |
| Unknown                                                                    | 44                            | 40                           | 4                        |                      |
| Activity as a precipitating factor                                         |                               |                              |                          | <0.01                |
| Yes                                                                        | 153 (51%)                     | 140 (58%)                    | 13 (23%)                 |                      |
| Unknown                                                                    | 44                            | 40                           | 4                        |                      |
| Joint type: large                                                          |                               |                              |                          | <0.01                |
|                                                                            | 144 (42%)                     | 88 (31%)                     | 56 (92%)                 |                      |
| Positive MTP/MCP squeeze test                                              |                               |                              |                          | <0.01                |
|                                                                            | 32 (9.4%)                     | 19 (6.8%)                    | 13 (21%)                 |                      |
| Normal bending of interphalangeal joints of the hands                      |                               |                              |                          | <0.01                |
|                                                                            | 326 (95%)                     | 275 (98%)                    | 51 (84%)                 |                      |
| Ability to make a fist                                                     |                               |                              |                          | <0.01                |
|                                                                            | 329 (96%)                     | 277 (99%)                    | 52 (85%)                 |                      |
| TipToe walking                                                             |                               |                              |                          | 0.03                 |
|                                                                            | 316 (95%)                     | 262 (96%)                    | 54 (89%)                 |                      |
| Unknown                                                                    | 8                             | 8                            | 0                        |                      |
| Presence of sacro-iliac tenderness                                         |                               |                              |                          | 0.11                 |
|                                                                            | 18 (5.3%)                     | 12 (4.3%)                    | 6 (9.8%)                 |                      |

JIA Juvenile idiopathic arthritis, MTP Metatarsophalangeal joints, MCP Metacarpophalangeal joints

<sup>1</sup>n (%)

<sup>2</sup>Wilcoxon rank sum test; Pearson's Chi-squared test; Fisher's exact test



**Fig. 1** Clinical items of the model. MTP: metatarsophalangeal joints; MCP: metacarpophalangeal joints; PIP: proximal interphalangeal joints; SI: sacroiliac. For the figures (Figure 1 and the easyJIA score in the supplementary material), we used the free online platform BeFunky (<https://www.befunky.com/about-us/>) to edit four photos (patient consent was provided) and in addition, we inserted an image created in BioRender in the last one: Chighizola, C. (2025) <https://BioRender.com/wr7gxhh>

### Simplified score

To improve usability in routine clinical practice, a simplified model was developed by rounding the regression coefficients to the nearest integer (see the “Rounded Estimate” column in Table 2). Negative coefficients were transformed into positive values by inverting the expected outcomes.

This resulted in a streamlined scoring system consisting of eight items (Fig. 3A), with a total possible score ranging from 0 to 10. The distribution of scores in our cohort is illustrated in Fig. 3B. The scoring system for daily practice can be found in the supplementary material. Each score corresponds to specific values for sensitivity, specificity, the predicted probability of JIA, positive predictive value (PPV), and negative predictive value (NPV), as detailed in Table 3. The ROC curve, representing the predicted risk of JIA based on the model’s regression score (calculated as the sum of the regression

coefficients multiplied by the values of the respective covariates), demonstrated an AUC of 0.92 with an overall accuracy of 0.88 (95% CI 0.84–0.91) using a cut-off of 3 points (Fig. 2B).

### Model internal validation

The model was internally validated using a bootstrap approach to create a validation dataset, onto which the developed model was applied. The resulting AUC was 0.92, with a 95% confidence interval (CI) ranging from 0.89 to 0.95 (Fig. 2C). Additionally, a comparison of the estimates and confidence intervals is presented in Fig. 2D.

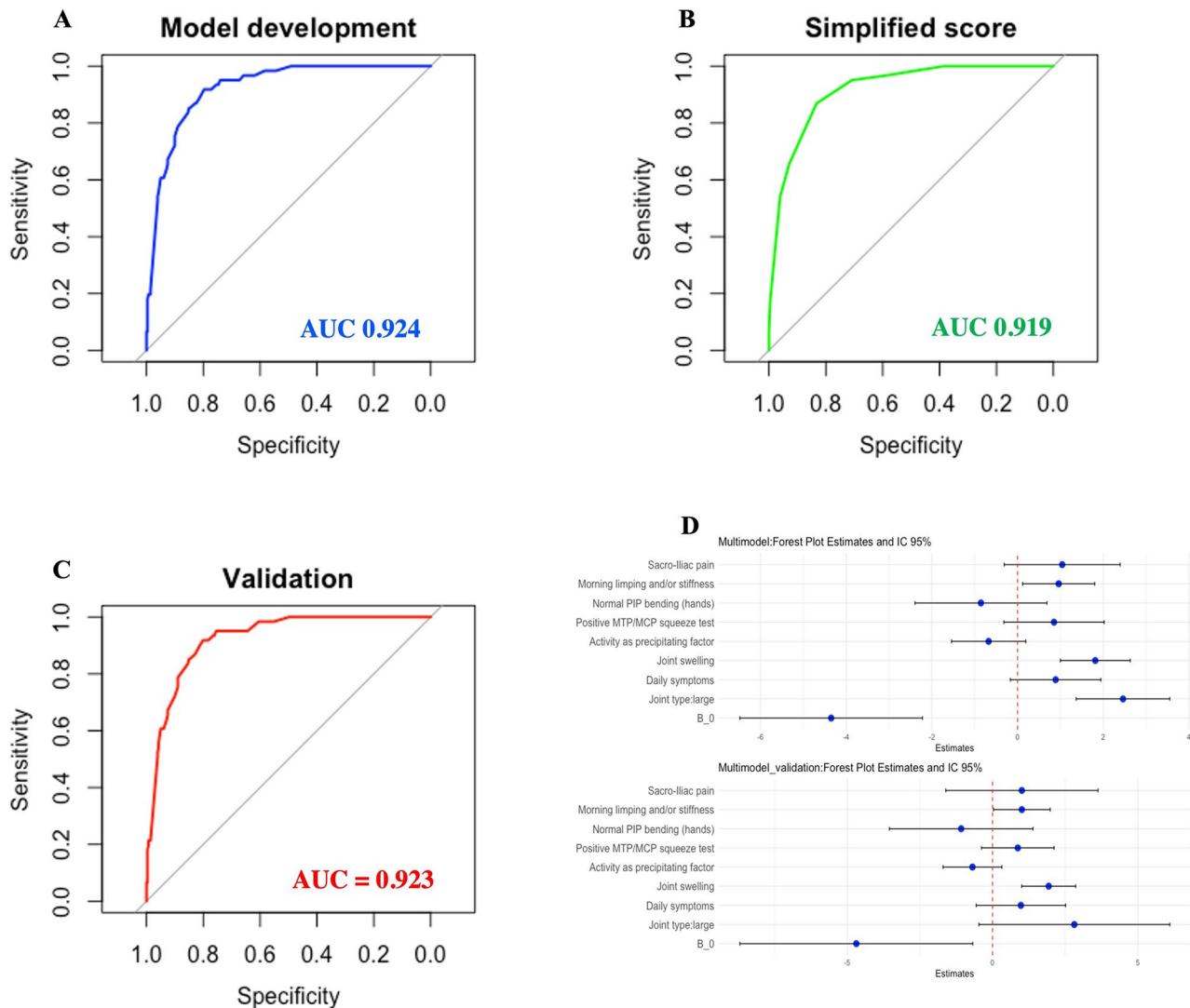
### Discussion

We present an evidence-based, simple, and time-efficient tool to assist clinicians in referring patients with suspected JIA. This scoring system also facilitates the screening of patients who may require further investigation, such as an eye exam, antibody testing or musculoskeletal ultrasound (MSK-US). The latter may help in daily clinics prioritizing patients who require specialized MSK-US assessments in centers with expertise in JIA US [14]. The items included in this score emphasize a more thorough characterization of joint complaints and incorporate simple maneuvers that do not require specialized training. Notably, the potential absence of joint swelling is expected to have only a limited impact on the overall score, which may help reduce the risk of overlooking patients with minimal joint enlargement who may benefit from a referral to pediatric rheumatology.

This tool is designed for use in primary care settings, where sensitivity should be prioritized. We therefore recommend using a cut-off of 3 or higher: this threshold provides high sensitivity (95%). In this regard, it must be highlighted that excluding from the table patients who are very unlikely to have JIA further emphasizes the high NPV as an indubitable advantage of the easyJIA. However, the score threshold could be adapted to the specific clinical setting, where communication between primary care and specialized centers is essential. To serve this scope, we provide the predicted probability of JIA, along with sensitivity, specificity, PPV, and NPV at each threshold. It should be noted that the frequency of the disease has a significant impact on both PPV and NPV.

We included sacroiliac tenderness in the scoring system to capture as many patients with enthesitis-related arthritis as possible, as these patients often experience the longest diagnostic delays—up to 15 months [8]. In our cohort, patients with ERA had a median score of 6, which corresponds to a predicted JIA probability of 50% and a PPV of 75%. To note, the lowest recorded score among ERA patients was 3.

A Brazilian group developed a rigorous instrument to identify children with clinical manifestations consistent



**Fig. 2** **A** ROC curve of the development model. **B** ROC curve of the simplified score with rounded estimates. **C** ROC curve of validation. **D** Estimates and 95% confidence intervals for the development model (top) and its validation (bottom) PIP: proximal interphalangeal joints; MTP: metatarsophalangeal joints; MCP: metacarpophalangeal joints

with chronic arthropathy. This questionnaire comprises 12 questions for patients or parents and demonstrates strong discriminatory ability. Indeed, a 50% probability of chronic arthropathy was associated with high values of sensitivity and specificity (90% and 93%, respectively). Nevertheless, it is important to note that this questionnaire was tested in patients with JIA, children with diffuse musculoskeletal pain, and healthy controls, which limits its discriminatory accuracy to these specific categories. Furthermore, this tool relies solely on self-reported assessments, with no input from clinicians or specialized healthcare providers, which may lead to the inclusion of many individuals who do not have JIA. Nonetheless, it appears valuable in contexts where the primary challenge is engaging individuals for their initial healthcare encounter [15]. In contrast, our scoring

system, while easy to implement, requires administration by a clinician or healthcare provider to mitigate potential screening issues of patients/parents self-reported assessment. Additionally, the design of our study, a diagnostic cohort study, offers a clear advantage in terms of the generalizability of the results compared to the case-control study conducted by the Brazilian group [15].

An Italian group developed a predictive score for chronic arthritis that also included patients presenting with fever [16]. Interestingly, no precipitating factors were found to be positively associated with a higher probability of chronic arthritis. Furthermore, compared to our scoring system, the pattern of joint swelling significantly influenced the final risk probability. This may impact the instrument's discriminatory capability, given the subjective nature of the reported information.

**Table 2** Logistic model presentation. MTP: metatarsophalangeal joints; MCP: metacarpophalangeal joints

| Variable                                              | Rounded Estimate | Estimate | 95% CI Lower | 95% CI Upper | P Value |
|-------------------------------------------------------|------------------|----------|--------------|--------------|---------|
| (Intercept)                                           | -4               | -4.35    | -6.48        | -2.22        | 0.00    |
| Joint type: large                                     | 2                | 2.46     | 1.37         | 3.55         | 0.00    |
| Daily symptoms                                        | 1                | 0.89     | -0.17        | 1.94         | 0.10    |
| Joint swelling                                        | 2                | 1.81     | 1.00         | 2.63         | 0.00    |
| Activity as precipitating factor                      | -1               | -0.68    | -1.54        | 0.19         | 0.13    |
| Positive MTP/MCP squeeze test                         | 1                | 0.85     | -0.32        | 2.02         | 0.16    |
| Normal bending of interphalangeal joints of the hands | -1               | -0.86    | -2.39        | 0.68         | 0.28    |
| Morning limping and/or stiffness                      | 1                | 0.96     | 0.12         | 1.80         | 0.03    |
| Presence of sacro-iliac tenderness                    | 1                | 1.04     | -0.31        | 2.39         | 0.13    |

MTP Metatarsophalangeal joints, MCP Metacarpophalangeal joints

JIA can have a gradual and insidious onset, often presenting with subtle yet progressive joint restriction that leads to functional impairment, particularly after prolonged periods of rest. This impairment may improve, sometimes completely, with movement, depending on the severity of joint involvement. In many cases, pain can be mild or even absent. Additionally, joint swelling can be difficult to detect due to factors such as the patient’s age, the specific joints affected, the degree of involvement, and the healthcare provider’s skill in joint examination. Limited confidence in musculoskeletal assessments presents another challenge [13]. Therefore, when evaluating a child with joint-related complaints, it is crucial to maintain a high level of suspicion for a JIA diagnosis, which often relies on the physician’s experience.

**Table 3** Measures corresponding to each score value

| Score | Sensitivity | Specificity | Predicted probability of JIA | PPV  | NPV  |
|-------|-------------|-------------|------------------------------|------|------|
| 0     | 1.00        | 0.19        | 0.00                         | 0.21 | 1.00 |
| 1     | 1.00        | 0.38        | 0.01                         | 0.26 | 1.00 |
| 2     | 0.97        | 0.59        | 0.02                         | 0.34 | 0.99 |
| 3     | 0.95        | 0.71        | 0.05                         | 0.41 | 0.98 |
| 4     | 0.87        | 0.83        | 0.12                         | 0.53 | 0.97 |
| 5     | 0.66        | 0.93        | 0.27                         | 0.67 | 0.93 |
| 6     | 0.54        | 0.96        | 0.50                         | 0.75 | 0.91 |
| 7     | 0.16        | 1.00        | 0.73                         | 1.00 | 0.85 |
| 8     | 0.07        | 1.00        | 0.88                         | 1.00 | 0.83 |
| 9     | 0.00        | 1.00        | 0.95                         | NaN  | 0.82 |
| 10    | 0.00        | 1.00        | 0.98                         | NaN  | 0.82 |

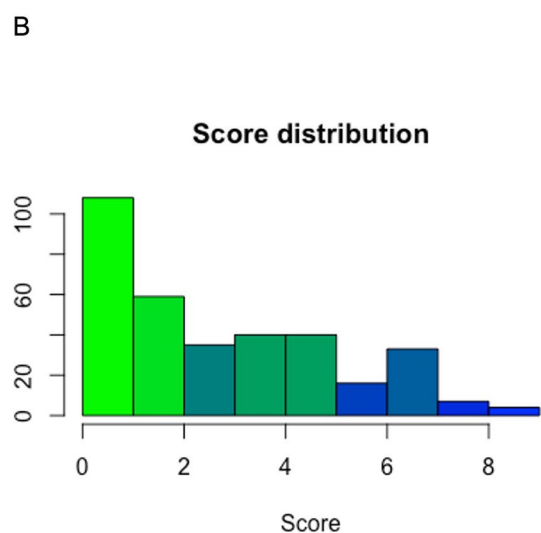
PPV Positive predictive value, NPV Negative predictive value, NaN Not a number

Not surprisingly, the median time from the first detected clinical manifestations to a JIA diagnosis remains between 4 and 5 months worldwide, with no clear trend toward reduction of such delay [8, 17].

Experiences from developed countries suggest that the accumulation of diagnostic delays often arises from the time taken to refer patients to PR after their initial encounter with healthcare providers [8, 9]. Prompt referral is essential, as evidence supports the concept that early diagnosis and treatment lead to better clinical outcomes [5, 6, 10, 11, 18]. Furthermore, early initiation of systemic treatments may have protective effects in patients with JIA-related uveitis by preventing local disease progression, reducing complications, and minimizing the need for topical steroids [19].

Similarly, there is substantial evidence supporting the benefit of early recognition and treatment of patients with rheumatoid arthritis (RA) [20–22]. The first structured effort to develop recommendations for the early

| Item                                                     | Potential points | Actual points |
|----------------------------------------------------------|------------------|---------------|
| Joint type: large                                        | 2                |               |
| Daily symptoms                                           | 1                |               |
| Joint swelling                                           | 2                |               |
| Activity as precipitating factor_no                      | 1                |               |
| Positive MTP/MCP squeeze test                            | 1                |               |
| Normal bending of interphalangeal joints of the hands_no | 1                |               |
| Morning limping and/or stiffness                         | 1                |               |
| Presence of sacro-iliac pain                             | 1                |               |



**Fig. 3** A the scoring system. B the score distribution in our cohort

referral of patients suspected of having RA dates back to the early 2000s, stemming from a systematic review combined with expert consensus [20]. Subsequently, further attempts have been made [23]. The experience of early arthritis clinics in adult rheumatology has further validated the benefits of early diagnosis and treatment [21, 22].

Several limitations affect this study. First, the setting: the research was conducted at a pediatric referral center rather than in a primary care office. Moreover, without external validation, it is difficult to confirm the robustness and reliability of the model across different contexts. Indeed, both the training and initial internal validation data may not fully capture the wide range of clinical scenarios that could occur in real-world settings, leading to spectrum bias. Although our findings are consistent with epidemiological data [24], it should also be acknowledged that the high frequency of Oligoarticular JIA observed in our cohort may influence the model's ability to screen for other JIA subcategories. Specifically, using fever as an exclusion criterion means our screening strategy is unlikely to identify systemic JIA/Still's disease; therefore, patients with fevers and arthralgia/arthritis should be referred urgently for in-hospital evaluation, with rheumatologic assessment considered as well, regardless of their screening easyJIA score. Another limitation, in certain settings, could be the adoption of a cutoff of 3 points, which tolerates a higher number of non-JIA referrals (false positives) compared to a cutoff of 4; however, we have provided all the necessary data to allow the choice of cutoff to be tailored to the characteristics of each work setting (Table 3). Additionally, this score has not been validated for referral of patients suspected of having systemic JIA/Still's disease. Although data missingness has been properly addressed under the assumption of MCAR/MAR, the mixed design of the study (retrospective and prospective enrollment) may limit the certainty of this notion.

On the other hand, the study has notable strengths. Excluding patients with fever results in a more homogeneous cohort, leading to more generalizable findings. The authors firmly believe that these patients warrant dedicated pathways; indeed, the presence of fever alongside arthralgias should be carefully investigated in an appropriate setting. The variable selection process using the bootstrap approach and LASSO technique reduces the risk of overfitting and enhances the model's adaptability to different settings. Furthermore, the combination of specific medical history details and straightforward maneuvers offers significant advantages. Indeed, this tool is intended for pediatricians and other healthcare providers who may encounter patients with suspected non-systemic JIA and likely do not require specialized training.

JIA diagnosis can be challenging primarily due to the absence of pathognomonic signs or definitive diagnostic tests. Therefore, early recognition of these patients and prompt treatment are crucial for achieving better outcomes. In this context, it is essential to promote educational programs at the primary care level and establish referral guidelines to facilitate access to pediatric rheumatology centers. Among the available tools for musculoskeletal assessment, the pGALS (paediatric Gait, Arms, Legs and Spine) should be mentioned as a simple and quick method for musculoskeletal screening in children [25]. Although not specifically designed as a screening tool for non-systemic JIA patients, its implementation during routine visits could further support physicians in differentiating normal from abnormal joints.

In this study, we presented and internally validated a simple, time-efficient scoring system designed to assist clinicians in the early referral of patients suspected of having non-systemic JIA. It is highly recommended to validate this score system in larger cohorts and within primary care settings. This would ensure its broader applicability, accuracy, and reliability in real-world clinical environments, while identifying potential limitations or areas for improvement to establish and potentially increase its overall validity.

### Supplementary Information

The online version contains supplementary material available at <https://doi.org/10.1186/s13075-025-03719-0>.

- Supplementary Material 1.
- Supplementary Material 2.
- Supplementary Material 3.
- Supplementary Material 4.

### Acknowledgements

The authors would like to express their heartfelt gratitude to Professor Rolando Cimaz for his guidance and inspiration.

### Authors' contributions

A.M., C.S., C.C., and R.C. conceived the study. A.M. drafted the manuscript. P.B., C.F., S.C., F.B., and M.G. were in charge of data acquisition. A.M., D.R., and C.S. were responsible for data analysis. All authors reviewed the manuscript, approved the submitted version, and agreed to be personally accountable for their contributions.

### Funding

No specific funding was received from any bodies in the public, commercial or not-for-profit sectors to carry out the work described in this article.

### Data availability

The data that support the findings of this study are available from the corresponding author, A.M., upon reasonable request.

## Declarations

### Ethics approval and consent to participate

Approval was granted by our center's Institutional Review Board (IRB) (4085\_S\_P). Informed consent for medical chart data collection was obtained from all patients or their legal guardians.

### Consent for publication

not applicable.

### Competing interests

The authors declare no competing interests.

Received: 11 July 2025 / Accepted: 19 December 2025

Published online: 09 January 2026

## References

1. Thierry S, Fautrel B, Lemelle I, Guillemin F. Prevalence and incidence of juvenile idiopathic arthritis: A systematic review. *Joint Bone Spine*. 2014;81(2):112–7.
2. Martini A, Lovell DJ, Albani S, Brunner HI, Hyrich KL, Thompson SD, et al. Juvenile idiopathic arthritis. *Nat Rev Dis Primers*. 2022;8(1):5.
3. Petty RE, Southwood TR, Manners P, Baum J, Glass DN, Goldenberg J, et al. International league of associations for rheumatology classification of juvenile idiopathic arthritis: second revision, Edmonton, 2001. *J Rheumatol*. 2004;31(2):390–2.
4. Weiss PF, Chauvin NA. Imaging in the diagnosis and management of axial spondyloarthritis in children. *Best Pract Res Clin Rheumatol*. 2020;34(6):101596.
5. Tynjälä P, Vähäsalo P, Tarkiainen M, Kröger L, Aalto K, Malin M, et al. Aggressive combination drug therapy in very early polyarticular juvenile idiopathic arthritis (ACUTE-JIA): A multicentre randomised open-label clinical trial. *Ann Rheum Dis*. 2011;70(9):1605–12.
6. Wallace CA, Giannini EH, Spalding SJ, Hashkes PJ, O'Neil KM, Zeff AS, et al. Trial of early aggressive therapy in polyarticular juvenile idiopathic arthritis. *Arthritis Rheum*. 2012;64(7):2012–21.
7. Chausset A, Pereira B, Echaubard S, Merlin E, Freychet C. Access to paediatric rheumatology care in juvenile idiopathic arthritis: what do we know? A systematic review. *Rheumatology (Oxford)*. 2020;59(12):3633–44.
8. Marino A, Baldassarre P, Ferrigno C, Biuso A, Minutoli M, Baldo F, et al. Pre-rheumatology referral consultation and investigation pattern in children with joint complaints: focus on juvenile idiopathic arthritis. *Child (Basel)*. 2024;11(5):600.
9. Shiff NJ, Abdwani R, Cabral DA, Houghton KM, Malleson PN, Petty RE, et al. Access to paediatric rheumatology subspecialty care in British Columbia, Canada. *J Rheumatol*. 2009;36(2):410–5.
10. Ravelli A, Martini A. Early predictors of outcome in juvenile idiopathic arthritis. *Clin Exp Rheumatol*. 2003;21(5 Suppl 31):93.
11. Sherry DD, Stein LD, Reed AM, Schanberg LE, Kredich DW. Prevention of leg length discrepancy in young children with pauciarticular juvenile rheumatoid arthritis by treatment with intraarticular steroids. *Arthritis Rheum*. 1999;42(11):2330–4.
12. Chausset A, Gominon AL, Montmaneix N, Echaubard S, Guillaume-Czitrom S, Cambon B, et al. Why we need a process on breaking news of juvenile idiopathic arthritis: A mixed methods study. *Pediatr Rheumatol Online J*. 2016;14:31.
13. Jandial S, Myers A, Wise E, Foster HE. Doctors likely to encounter children with musculoskeletal complaints have low confidence in their clinical skills. *J Pediatr*. 2009;154(2):267–71.
14. Cimaz R, Giani T, Caporali R. What is the real role of ultrasound in the management of juvenile idiopathic arthritis? *Ann Rheum Dis*. 2020;79(4):437–9.
15. Len CA, Terreri MT, Puccini RF, Wechsler R, Silva EK, Oliveira LM, et al. Development of a tool for early referral of children and adolescents with signs and symptoms suggestive of chronic arthropathy to pediatric rheumatology centers. *Arthritis Rheum*. 2006;55(3):373–7.
16. Cattalini M, Parissenti I, Tononcelli E, Lancini F, Cantarini L, Meini A. Developing a predictive score for chronic arthritis among a cohort of children with musculoskeletal complaints—The chronic arthritis score study. *J Pediatr*. 2016;169:188–93.
17. McErlane F, Foster HE, Carrasco R, Baildam EM, Chieng SEA, Davidson JE, et al. Trends in paediatric rheumatology referral times and disease activity indices over a ten-year period among children and young people with juvenile idiopathic arthritis: results from the childhood arthritis prospective study. *Rheumatology (Oxford)*. 2016;55(7):1225–34.
18. Ong MS, Ringold S, Kimura Y, Schanberg LE, Tomlinson GA, Natter MD, CARRA Registry Investigators. Improved disease course associated with early initiation of biologics in polyarticular juvenile idiopathic arthritis: trajectory analysis of a childhood arthritis and rheumatology research alliance consensus treatment plans study. *Arthritis Rheumatol*. 2021;73(10):1910–20.
19. van Straalen JW, Akay G, Kouwenberg CV, de Roock S, Kalinina Ayuso V, Wulffraat NM, et al. Methotrexate therapy associated with a reduced rate of new-onset uveitis in patients with biological-naïve juvenile idiopathic arthritis. *RMD Open*. 2023;9(2):e003010.
20. Emery P, Breedveld FC, Dougados M, Kalden JR, Schiff MH, Smolen JS. Early referral recommendation for newly diagnosed rheumatoid arthritis: evidence based development of a clinical guide. *Ann Rheum Dis*. 2002;61(4):290–7.
21. Verhoeven MM, de Hair MJ, Tekstra J, Bijlsma JW, van Laar JM, Pethoe-Schramm A, et al. Initiating tocilizumab, with or without methotrexate, compared with starting methotrexate with prednisone within step-up treatment strategies in early rheumatoid arthritis: an indirect comparison of effectiveness and safety of the U-Act-Early and CAMERA-II treat-to-target trials. *Ann Rheum Dis*. 2019;78(10):1333–8.
22. Nisar MK. Early arthritis clinic is cost-effective, improves outcomes and reduces biologic use. *Clin Rheumatol*. 2019;38(6):1555–60.
23. van Delft ETAM, Barreto DL, van der Helm-van Mil AHM, Alves C, Hazes JMW, Kuijper TM, et al. Diagnostic performance and clinical utility of referral rules to identify primary care patients at risk of an inflammatory rheumatic disease. *Arthritis Care Res (Hoboken)*. 2022;74(12):2100–7.
24. Consolaro A, Giancane G, Alongi A, van Dijkhuizen EHP, Aggarwal A, Al-Mayouf SM, Paediatric Rheumatology International Trials Organisation, et al. Phenotypic variability and disparities in treatment and outcomes of childhood arthritis throughout the world: an observational cohort study. *Lancet Child Adolesc Health*. 2019;3(4):255–63.
25. Foster HE, Jandial S. pGALS - paediatric gait arms legs and spine: a simple examination of the musculoskeletal system. *Pediatr Rheumatol Online J*. 2013;11(1):44.

## Publisher's note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.