

Prevalence and complications of uveitis in juvenile idiopathic arthritis in a population-based nation-wide study in Germany: suggested modification of the current screening guidelines

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Objectives. To analyse the prevalence and complications of uveitis and their predictors in a large cohort of patients with juvenile idiopathic arthritis (JIA).

Methods. Data of 3271 JIA patients as classified by International League of Associations for Rheumatology (ILAR) criteria included in a national database during 1 yr were analysed.

Results. Uveitis prevalence was 12% of all JIA patients. The most frequent were oligoarthritis extended (25%) and persistent (16%). JIA patients with uveitis were significantly younger at onset of arthritis (3.8 vs 7.0 yrs) or ANA-positive (86% vs 42%) than the patients without uveitis. Predictors of uveitis included age at onset ($P=0.03$) and ANA-positivity ($P<0.01$) besides the presence of a certain JIA subgroup ($P=0.04$). Uveitis was clinically silent in 75% of the oligoarthritis but in none of the enthesitis-related arthritis patients. The median onset of uveitis was 5.5 months after arthritis manifestation. In 73%, 77% and 90%, uveitis developed within 1, 2 and 4 yrs after arthritis, respectively. Anterior uveitis was the most common anatomic type of uveitis (83%). Uveitis complications at mean follow-up of 5.6 yrs were common (56%), and predictors for complications included presence of complications at first visit ($P<0.001$) and uveitis manifestation before arthritis ($P=0.001$), but not ANA positivity.

Conclusions. The JIA subgroups markedly differ with respect to the prevalence and course of associated uveitis. Ophthalmological screening should be initiated early after arthritis onset and the intervals be related to the JIA subgroup. A modification of the current screening guidelines is suggested.

KEY WORDS: Uveitis, Iridocyclitis, Prevalence, Juvenile idiopathic arthritis, National paediatric rheumatological database.

Introduction

Juvenile idiopathic arthritis (JIA) is a heterogeneous group of chronic arthritis diseases in childhood. JIA is frequently accompanied by a vision-threatening uveitis [1, 2]. Previously, the prevalence of uveitis has been found to be between 4% and 24.4% [3, 4].

The confusingly different prevalence rates can be attributed to the different study designs used. The previous studies were based on different classification criteria for childhood chronic arthritis, e.g. the American College of Rheumatology (ACR) or the European League Against Rheumatism (EULAR) criteria [5, 6]. There are still no large investigations available using the currently applied International League of Associations for Rheumatology (ILAR) criteria. The few population-based data are of limited value due to the small numbers of patients included [7].

The studies from tertiary uveitis and rheumatology centres were profoundly different owing to selection criteria and follow-up.

However, accurate estimation of uveitis prevalence and complications is mandatory for the optimization of screening guidelines for uveitis in JIA patients [1, 2]. It is of importance that these guidelines refer to the ACR classification of arthritis and that there are still no guiding principles provided for the ILAR classification. And furthermore, the previous studies that determined the ocular complications or visual loss mostly have not been population-based, did not specifically address the subgroup of children with juvenile idiopathic arthritis, and included various infectious and non-infectious diseases [8–12].

Most recently, a nationwide registry on children and adolescents with juvenile rheumatic diseases was established. Herein, we present the data of 3271 JIA cases documented by paediatric rheumatologists and ophthalmologists during 1 yr.

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Materials and methods

Clinical data of children and adolescents with JIA have been recorded once a year by paediatric rheumatologists within the national paediatric rheumatological database. In 2002, a uveitis-questionnaire was added to the rheumatological database that inquired eye findings from JIA patients with uveitis. This questionnaire was given to all JIA patients with established uveitis and filled out by their treating ophthalmologists.

Subjects' consent was obtained according to the Declaration of Helsinki. The design of the work conforms to the standards currently applied in Germany and has been approved by the ethics committee in Berlin.

All JIA cases recorded between 1 January 2002 and 31 December 2002 by tertiary paediatric rheumatology centres and ophthalmological practices as well were considered for this analysis [13] (Fig. 1). All patients included herein fulfilled the ILAR criteria for JIA [14]. The paediatric rheumatological

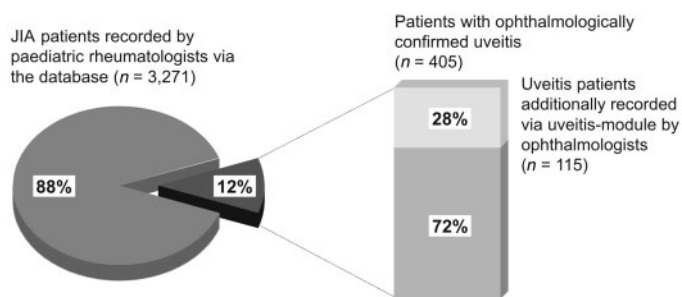


FIG. 1. All patients with JIA recorded by tertiary paediatric rheumatology centres and ophthalmological practices. National database in Germany during the year 2002.

database included age, gender, arthritis subgroup, age at onset of arthritis, treatment, arthritis activity, number of swollen or tender joints and/or joints with limited range of motion (ROM), extraarticular manifestations such as the presence of uveitis and uveitis complications. The presence of antinuclear antibodies (ANA), HLA-B27 antigen and rheumatoid factor (RF) were documented.

The parameters included in the ophthalmological database were age, gender, age at onset of uveitis, uni- or bilateral uveitis, initial and current best-corrected visual acuity, anti-inflammatory therapy for uveitis, initial and current eye complications and previous eye surgery. Uveitis was anatomically classified in anterior, intermediate, posterior and pan-uveitis and is in agreement with the most recently provided uveitis standardization [15]. Symptomatic uveitis with sudden onset of redness and pain was differentiated from asymptomatic, clinically silent uveitis.

All of the available data from the rheumatological and ophthalmological database were analysed at the German Rheumatism Research Center in Berlin (DRFZ). Data analysis was achieved with SPSS 11.0 statistical package under Windows 2000 (Norusis 2002: SPSS Advanced Statistics 11.0. Chicago SPSS Inc., IL, USA). The chi-square test for categorical data and Student's *t*-test were used for statistical analysis when appropriate. Logistic regression analyses were applied to identify predictors of uveitis and its complications at documentation. Results were expressed as odds ratios (ORs) and confidence intervals (CIs). A significance level of 5% was used for all analyses.

Results

Data collected from 3271 patients out of 35 centres were included in this analysis. Uveitis was noted in 406 (12%) of the cases. Ophthalmological data were documented in 115 of the uveitis patients (28%). It is noteworthy that the data from the 115 patients with documented course of uveitis did not differ from the other uveitis patients with respect to arthritis activity, joint function, gender, duration of arthritis and therapy.

The majority of all JIA patients recorded had oligoarthritis, followed by seronegative (RF negative) polyarthritis and enthesitis-related arthritis (ERA). While uveitis was particularly common in patients with oligoarthritis, it was rare in patients with systemic arthritis and RF-positive polyarthritis (Table 1). The different uveitis risks of the various JIA subgroups were disclosed by univariate analysis showing for example a 20- and 30-fold higher uveitis risk in persistent and extended OA, respectively, as compared with systemic arthritis (Table 2).

The characteristics of JIA patients with and without documented uveitis were compared (Table 3). Uveitis occurred between 0 and 16 yrs of age, with a mean of 5.2 yrs (s.d. 3.2). Approximately three in four patients (77%) developed uveitis before their 7th birthday. Patients with uveitis were significantly younger at onset of arthritis, more often female and ANA-positive than the JIA patients without uveitis. In contrast to the

TABLE 1. Cumulative uveitis incidence in the different juvenile idiopathic arthritis (JIA) subgroups after median duration of 4.3 yrs

JIA subgroup	No. of patients with uveitis	Cumulative uveitis incidence, % (95% CI)	Median disease duration, yrs (IQR)
Systemic arthritis	2/198	1 (0–2.4)	4 (5)
Oligoarthritis (persistent)	246/1497	16 (14.6–18.3)	3 (5)
Oligoarthritis (extended)	57/227	25 (19.5–30.7)	6 (6)
Polyarthritis (RF negative)	19/405	4 (2.6–6.8)	4 (5)
Polyarthritis (RF positive)	3/67	2 (0–9.4)	4 (3)
Enthesitis-related arthritis	26/384	7 (4.2–9.3)	3 (3)
Psoriatic arthritis	26/251	10 (6.6–14.1)	3 (5)
Other arthritis	27/242	11 (7.2–15.1)	3 (4)
All JIA patients	406/3271	12 (11.3–13.5)	3 (5)

RF = rheumatoid factor, IQR = interquartile range.

TABLE 2. Uveitis in juvenile idiopathic arthritis (JIA) subgroups in a population-based nation wide study in Germany

JIA subgroups	<i>P</i> -value	OR (95% CI)
Systemic arthritis		1
Oligoarthritis (persistent)	<0.01	19 (4.7–78.1)
Oligoarthritis (extended)	<0.01	33 (7.9–136.6)
Polyarthritis (RF negative)	0.036	5 (1.1–20.9)
Polyarthritis (RF positive)	0.275	3 (0.4–21.8)
Enthesitis-related arthritis	0.008	7 (1.7–30.3)
Psoriatic arthritis	0.001	11 (2.7–48.3)
Other arthritis	0.001	12 (2.9–52.4)

Predictors of uveitis by univariate analysis.

TABLE 3. Uveitis in juvenile idiopathic arthritis (JIA) subgroups in a population-based nation wide study in Germany

	JIA patients without uveitis	JIA patients with uveitis	<i>P</i> -value
Number of patients	2865 (%)	115 (%)	
JIA subgroups, <i>n</i> (%)			<i>P</i> < 0.001
Systemic arthritis	198 (7)	0	
Oligoarthritis (persistent)	1251 (44)	71 (62)	
Oligoarthritis (extended)	170 (6)	20 (17)	
Polyarthritis (RF negative)	386 (14)	7 (6)	
Polyarthritis (RF positive)	65 (2)	0	
Enthesitis-related arthritis	358 (13)	4 (3)	
Psoriatic arthritis	225 (8)	4 (3)	
Other arthritis	215 (8)	9 (8)	
Disease onset in yrs, mean (s.d.)	7.0 (4.1)	3.8 (2.8)	<i>P</i> < 0.001
Duration of disease in yrs, mean (s.d.)	4.0 (3.6)	6.5 (4.1)	<i>P</i> = 0.002
Female (%)	62	74	<i>P</i> = 0.012
ANA-positive (%)	42	86	<i>P</i> < 0.001

oligoarthritis and RF-negative polyarthritis patients with uveitis, the ERA patients commonly were HLA-B27 positive (75%) and were male (75%). By multivariate analysis, predictors of uveitis included age at onset (*P* = 0.03, OR 0.95, CI 0.90–0.99), disease duration (*P* < 0.01, OR 1.11; CI 1.07–1.15) and ANA-positivity (*P* < 0.01; OR 2.63, CI 1.83–3.79) besides the presence of a certain JIA subgroup (*P* = 0.04).

The predictors of uveitis were additionally examined for the group of early cases (disease duration <24 months) with oligoarthritis (*n* = 330) that appeared to be at a particularly high risk of uveitis. As determined by multivariate analysis, again ANA-positivity (*P* < 0.001; OR 10.7, CI 3.1–37.1) was a predictor of uveitis, whereas HLA-B27 (*P* = 0.06), age at onset (*P* = 0.42), gender (*P* = 0.23), number of joints with arthritis (*P* = 0.38) and ESR (*P* = 0.08) had no significant influence.

The median onset of uveitis was 5.5 months (mean 21) after arthritis manifestation for the entire study population. Uveitis occurred before the onset of arthritis in 10% and these patients had asymptomatic uveitis. Uveitis appeared simultaneous with or within the first 6 months of arthritis onset in 48% and between

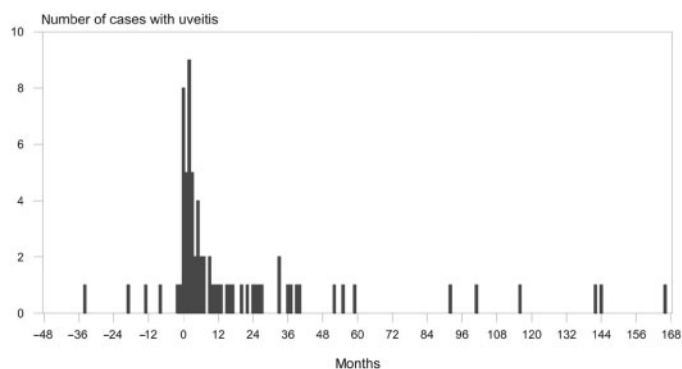


FIG. 2. Onset of uveitis before or after onset of arthritis. National database in Germany during the year 2002.

TABLE 4. Uveitis in juvenile idiopathic arthritis (JIA) subgroups in a population-based nation wide study in Germany

JIA subgroup	Complications ^a n (%)	Visual acuity ^b		Asymptomatic uveitis n (%)
		≤20/200 n (%)	≤20/50 n (%)	
Oligoarthritis (persistent)	36 (54)	13 (9)	42 (30)	62 (87)
Oligoarthritis (extended)	11 (58)	9 (23)	15 (38)	7 (78)
Polyarthritis (RF negative)	4 (67)	1 (7)	3 (21)	7 (100)
Enthesitis-related arthritis	0 (0)	2 (25)	4 (50)	1 (33)
Psoriatic arthritis	1 (25)	0	1 (13)	3 (75)
Other arthritis	7 (87)	6 (33)	8 (44)	2 (40)
All patients	59 (56)	31 (14)	73 (32)	85 (80)

Uveitis complications and visual acuity at final documentation, and clinically silent (asymptomatic) uveitis. ^an=106 patients; ^bn=226 eyes.

6 and 24 months in 19%. Taken together, 73% of patients exhibited uveitis before or within the first 12 months after onset of arthritis and 77% and 90% within the first 2 and 4 yrs, respectively. Uveitis manifestation was between 2 and 5 yrs and 5 yrs and more after arthritis manifestation in 16% and 7%, respectively (Fig. 2). In all patients with arthritis onset >6 yrs (15% of the uveitis patients), the uveitis developed within the following 2 yrs.

In the ERA patients, two out of three had symptomatic relapses of uveitis. In contrast, uveitis was asymptomatic in most of the oligoarthritis and RF-negative polyarthritis patients (Table 4).

Whereas anterior uveitis was the most common anatomic type of uveitis (83%), intermediate uveitis (9%), posterior uveitis (1%) and pan-uveitis (7%) were less frequent. In 70% of all uveitis patients, the uveitis was bilateral. Both eyes were involved in 77% of the cases with oligoarthritis.

In 45% of the patients, uveitis complications were already noted at the initial visit by the currently treating ophthalmologist. Altogether, uveitis was diagnosed an average of 7 months after this initial eye examination.

In 56%, uveitis complications were present at the final ophthalmological documentation at a mean of 5.6 yrs (s.d. 4.9) after onset of uveitis (Table 4). Specifically, the complications at initial and final status are compared in Table 5. A vision of ≤20/200 or 20/50 was detected in at least one eye in 15% and 46%, respectively.

By using univariate logistic regression analysis, the following predictors of complications at the final visit were determined: presence of complications on the first visit at the currently treating ophthalmologist ($P < 0.001$, OR 80.2, CI 16.7–383.9) and manifestation of uveitis before arthritis ($P = 0.001$, OR 20.8, CI 2.5–171.4). In contrast, JIA subgroup, ANA positivity, gender, age at onset and disease duration were not found to be of significant influence. Thirty-two per cent of the patients required

TABLE 5. Number of patients with ocular complications due to uveitis

	Initial status (n=100) n (%)	Current status (n=106) n (%)
Posterior synechia	31 (31)	29 (27)
Cataract	22 (22)	27 (26)
Band-keratopathy	21 (21)	31 (29)
Vitreous opacities	10 (10)	7 (7)
Papille oedema	7 (7)	6 (6)
Secondary glaucoma	6 (6)	8 (8)
Macula oedema	4 (4)	6 (6)
Hypotony	3 (3)	4 (4)
Iris rubeosis	2 (2)	2 (2)
Other complications	9 (9)	22 (21)

eye surgery for the uveitis complications. The most frequent operations were cataract surgery (29%), pars-plana vitrectomy (10%) and glaucoma surgery (4%).

Discussion

The 3271 patients included in this documentation represent at least one-fifth of the expected number of JIA patients in Germany, assuming a JIA prevalence of ~0.1%. The JIA subgroup distribution of this patient cohort was comparable with that of other population-based studies, which underlines that the data are representative. The fact that ophthalmological data were documented in only 115 of the uveitis patients (28%) may limit the analysis. Data of the rheumatological database suggest that the uveitis patients recorded by ophthalmologists had more often uveitis sequelae (42%) than those not recorded (29%, $P = 0.02$). However, as the uveitis patients for whom ophthalmological data were available did not differ from those not recorded by ophthalmologists with respect to age at onset, gender, JIA subgroup or disease severity, age at onset and length of uveitis, the observations respective to the uveitis incidence are also supposed to be representative. In our study, 12% out of the 3271 JIA patients had eye involvement.

Previously, the American Section of Rheumatology and Ophthalmology has provided guidelines for the routine screening for uveitis in juvenile rheumatoid arthritis (JRA) patients that referred to the ACR criteria [1, 2]. It is noteworthy that there are currently no screening guidelines provided that applied the ILAR classification.

This is the largest population-based epidemiological study to date on uveitis in JIA patients that used the current ILAR classification. Although the observations herein confirm in many respects the previous screening guidelines [1, 2], certain modifications appear to be reasonable. Also, strategies are introduced for subgroups of arthritis patients that have not been mentioned in the previous recommendations. Specifically, unclassified arthritis frequently associated with uveitis in our registry and patients with psoriatic arthritis and ERA are included.

The observations herein disclose that the risk of uveitis differs enormously between the various JIA subgroups. The vast majority of uveitis patients in the study herein had oligoarthritis (79%). This number is in agreement with previous observations [2, 16]. Our data support the high frequency of ANA in oligoarthritis-associated uveitis (86%) that was also noted previously [4, 17, 18]. The observations herein furthermore disclose that other subgroups are also frequently associated with uveitis, as psoriatic arthritis, unclassified arthritis and ERA.

Similar to previous reports, uveitis occurred predominantly in JIA children with early onset of disease in our study [3, 17]. Herein, mean arthritis onset was 7.0 yrs (median 7.0, range 0–15) in JIA patients without uveitis and 3.8 yrs (median 3.0, range 0–15) in patients who had uveitis. The mean age of uveitis manifestation in JIA patients was 7 yrs in previous studies [19]

and 5.2 yrs (median 4, range 0–16) in this study. However, the observations herein illustrate that uveitis may occur at any age.

In only 2.7–6.8% of the JIA patients, the uveitis manifests itself before arthritis [3, 17, 19, 20], in our study in 10% of the children. It has been previously shown that in 25 to 33% of the patients, uveitis was detected synchronously with the onset of arthritis [3, 17]. In ~50%, it appeared within the first 3 months after arthritis [4, 21], and in 90% within the first 4 yrs [4], but may occasionally develop up to 10 yrs after arthritis [22]. In our study, uveitis appeared before or within the first 12 months after onset of arthritis in 73% of the patients. In ~5% of patients, uveitis manifested between 2 and 4 yrs after arthritis onset and in 7% even after 5 or more years. In agreement with the previous guidelines [1, 2], therefore, screening for uveitis should be instituted immediately with the diagnosis of JIA and on the basis of the data, must be continued for 7 yrs. In accordance with the previous guidelines, yearly screenings may be continued indefinitely in all groups.

Additionally, the data herein illustrate that clinically silent uveitis in a white eye is typical for oligoarthritis, RF-negative polyarthritis, unclassified arthritis and in psoriatic arthritis, especially with early-onset (data not shown). This points out the need for short-interval ophthalmological evaluations in these JIA subgroups. In contrast, the screening intervals may be longer in HLA-B27-positive ERA patients [3, 16], as uveitis is symptomatic in these patients. Generally, a symptomatic uveitis relapse may prompt an ophthalmic visit. However, yearly visits may be performed in patients that are at high risk of such uveitis, as symptoms may not be obvious and complications may arise. This may be useful for patients in subgroups with HLA-B27-positive ERA or late-onset psoriatic arthritis.

High complication rates have been previously reported for JIA uveitis [8–12, 19]. Notably, up to one-third of the patients had already presented with complications at the time of uveitis diagnosis [4, 11, 17]. The more recently published papers indicated that the incidence of complications is currently lower. However, the proportion of JIA patients with uveitis complications remains higher than in other types of uveitis.

Interestingly, the proportion of patients with complications was already 45% at initial presentation to the documenting ophthalmologist in our study and as high as 56% at the time of documentation. The high percentages of ocular complications of paediatric uveitis patients have also been noted most recently by others [8]. The high number may result from the fact that the presence of any of the uveitis complications was counted. It is noteworthy in this respect that the mean time since diagnosis of uveitis herein was 5.6 yrs at the time of documentation. Additionally, the patients were not routinely registered at the initial presentation of uveitis and the earliest findings were not always available for registration. The high frequency of JIA uveitis patients in our study group who required immunosuppression (data not shown) or eye surgery, moreover, documents the severity of this type of uveitis, although the patients did not come predominantly from uveitis centres.

Our observations herein suggest that poor vision is generally related to the presence of uveitis complications. It has been previously shown that poor initial vision was more common when uveitis occurred before or at the time of arthritis onset [20, 21] and when complications had already been detected at the first presentation. In our study, manifestation of uveitis before arthritis has also been a strong predictor for uveitis complications. Therefore, early diagnosis appears to be especially important and may ameliorate the outcome profoundly.

As a consequence, a 3-month screening interval is herein suggested for those JIA subgroups with an asymptomatic course and a high frequency of uveitis and complications (Table 6). Similar to previous publications, the data with reference to uveitis complications and symptoms is limited by the number of patients

TABLE 6. Suggested screening intervals for uveitis in patients with JIA as classified by International League of Associations for Rheumatology (ILAR) criteria

JIA subgroup	ANA	Age at JIA onset (yrs)	JIA duration (yrs)	Recommended screening intervals (months)
OA, RF-PA, PsA, AA	+	≤6	≤4	3
OA, RF-PA, PsA, AA	+	≤6	>4	6
OA, RF-PA, PsA, AA	+	≤6	≥7	12
OA, RF-PA, PsA, AA	+	>6	≤2	6
OA, RF-PA, PsA, AA	+	>6	>2	12
OA, RF-PA, PsA, AA	-	≤6	≤4	6
OA, RF-PA, PsA, AA	-	≤6	>4	12
OA, RF-PA, PsA, AA	-	>6	n.a.	12
ERA	n.a.	n.a.	n.a.	12
RF-PA, Sys A	n.a.	n.a.	n.a.	12
Patients with uveitis	n.a.	n.a.	n.a.	According to uveitis course

Sys A = systemic arthritis; OA = oligoarthritis; RF-PA = seronegative polyarthritis; RF + PA = seropositive polyarthritis; ERA = enthesitis-related arthritis; PsA = psoriatic arthritis; AA = other arthritis; n.a. = not applicable.

included in this documentation. However, the recommendations herein are supported by the published findings.

It has been previously described that patients with psoriatic arthritis with an onset of arthritis <4 yrs are also at a high risk for a chronic type of asymptomatic uveitis [23]. As this is supported by our findings, screening intervals should be applied that are similar to the oligoarthritis subgroup.

The typical complications published previously included cataract (19–81%), glaucoma (8–38%), band keratopathy (7–70%), posterior synechiae (8–75%) and ocular hypotony (19%) [3, 4, 8, 10, 16, 19, 24]. Similar numbers were also found in our study. As macular oedema was frequently noted in the previous and also the present study [17, 20], beside slit-lamp examination, funduscopy should always be considered.

It has been repeatedly shown that anterior uveitis is characteristic for JIA patients and our data (87%) are in agreement with this notion. Principally, the uveitis classification used herein was based on the anatomic location of uveitis and is in accordance with the most recently published standardization [15]. However, the participating ophthalmologists classified the uveitis in some of the JIA patients as intermediate (9%) or posterior (1%) uveitis. We believe that in some of these patients this may be a misclassification due to the presence of structural complications, e.g. cell infiltration in the vitreous or macular oedema that are also typical complications from iridocyclitis.

While ANA is a strong predictor for uveitis, complications in ANA-negative JIA patients were as common as in the ANA-positive children. Furthermore, our patients with oligoarthritis or polyarthritis onset being ANA negative frequently showed an asymptomatic course of uveitis. Screening intervals in the ANA-negative patients may also be shorter than in the previous guidelines [1, 2] in order to avoid a delay of the recognition and treatment of recurrence. This recommendation is in accordance with the observations by Chalom and colleagues [25] that complications were more common in ANA-negative than in ANA-positive patients.

The prospective population-based documentation may facilitate the optimization of the current guidelines for the screening intervals and thus may help to improve the long-term outcome of these patients.

Rheumatology key messages

- JIA subgroups differ with respect to prevalence and course of uveitis.
- Modified screening is provided for uveitis in JIA as classified by ILAR criteria.

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