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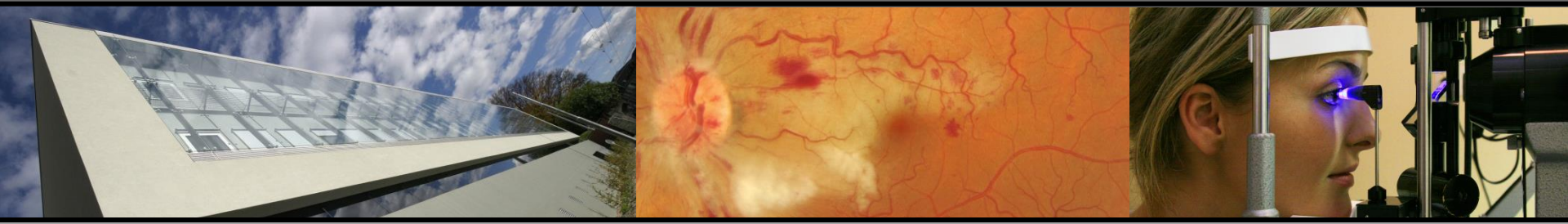
Jules-Gonin
Service universitaire d'ophtalmologie
Fondation Asile des aveugles

UNIL | Université de Lausanne
Faculty of Biology and Medicine



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Uveitis in children and JIR cohorte

Privat Docent, MER-1

Médecin Adjoint
Immuno-infectiologie oculaire
Département d'ophtalmologie
Hôpital Jules-Gonin
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Plan

- SUN classification
- Epidemiology
- JIA and complications
- Herpetic kerato-uveitis
- Pars planitis
- Toxoplasmosis / CMV
- JIR cohort and conclusions

PERSPECTIVES

Standardization of Uveitis Nomenclature for Reporting Clinical Data. Results of the First International Workshop

THE STANDARDIZATION OF UVEITIS NOMENCLATURE (SUN) WORKING GROUP

Am J Ophthalmol 2005;140: 509-516

Standardiz ture for
Reporting ne First

THE STANDAR G GROUP

l 2005;140: 509-516

TABLE 1. The SUN* Working Group Anatomic Classification of Uveitis

Type	Primary Site of Inflammation†	Includes
Anterior uveitis	Anterior chamber	Iritis
		Iridocyclitis
		Anterior cyclitis
Intermediate uveitis	Vitreous	Pars planitis
		Posterior cyclitis
		Hyalitis
Posterior uveitis	Retina or choroid	Focal, multifocal, or diffuse choroiditis
		Chorioretinitis
		Retinochoroiditis
		Retinitis
		Neuroretinitis
Panuveitis	Anterior chamber, vitreous, and retina or choroid	

*SUN = Standardization of uveitis nomenclature.
†As determined clinically. Adapted from the International Uveitis Study Group anatomic classification in reference 1.

TABLE 2. The SUN* Working Group Descriptors of Uveitis

Category	Descriptor	Comment
Onset	Sudden	
	Insidious	
Duration	Limited	≤3 months duration
	Persistent	>3 months duration
Course	Acute	Episode characterized by sudden onset and limited duration
	Recurrent	Repeated episodes separated by periods of inactivity without treatment ≥3 months in duration
	Chronic	Persistent uveitis with relapse in <3 months after discontinuing treatment

*SUN = Standardization of uveitis nomenclature.

TABLE 3. The SUN* Working Group Grading Scheme for Anterior Chamber Cells

Grade	Cells in Field†
0	<1
0.5+	1–5
1+	6–15
2+	16–25
3+	26–50
4+	>50

*SUN = Standardization of uveitis nomenclature.
†Field size is a 1 mm by 1 mm slit beam.

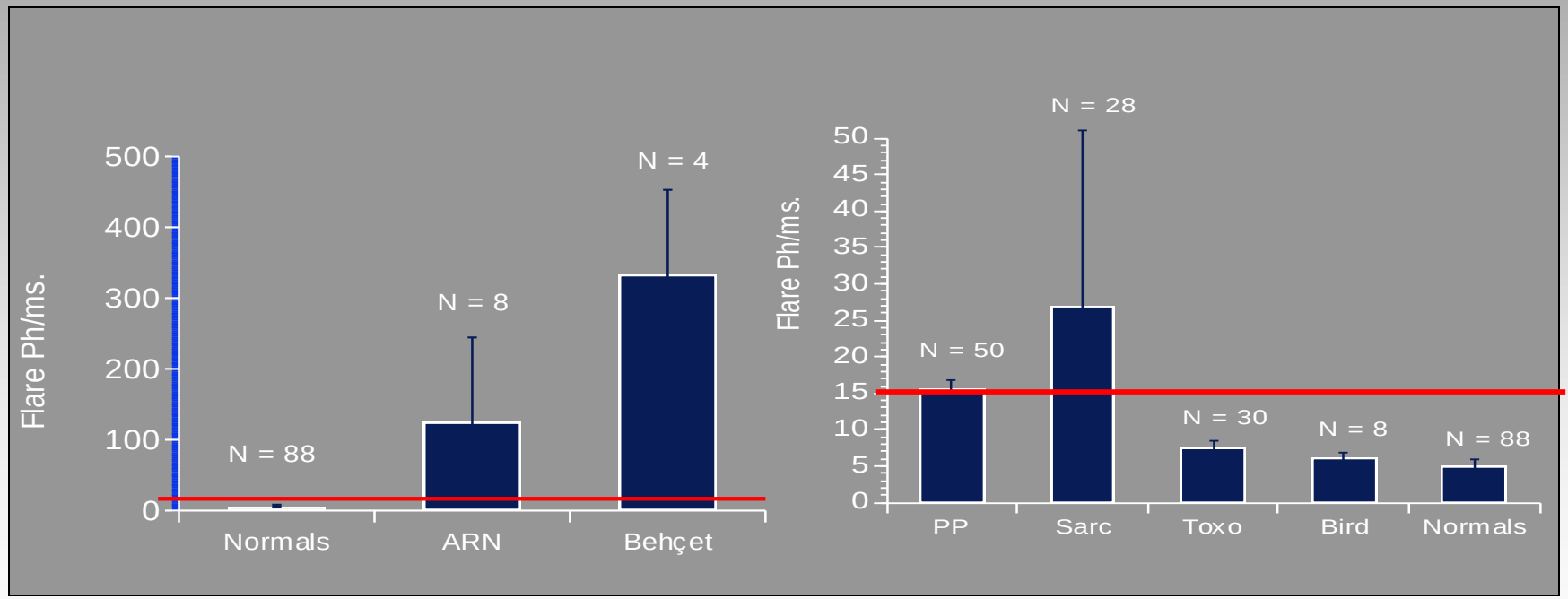
TABLE 4. The SUN* Working Group Grading Scheme for Anterior Chamber Flare

Grade	Description
0	None
1+	Faint
2+	Moderate (iris and lens details clear)
3+	Marked (iris and lens details hazy)
4+	Intense (fibrin or plastic aqueous)

Adapted from reference 12.
*SUN = Standardization of uveitis nomenclature.

Laser flare photometry in posterior uveitis

Guex-Crosier Ophtalmology 1994



Uveitis in children

- 5 – 10 % of adult uveitis (145 children / 1540 Uveitis followed at Jules Gonin Eye Hospital)
- Incidence 4.9 – 6.9 / 100 000 children / year
- Prevalence 13 – 30 / 100 000 children
- Legal blindness 17 – 23 % of children
- AJI : Probability of 45% in the presence of bilateral UAA
- AJI : Proabability of 35% in the presence of unilateral UAA
  - 36 – 67 % of cases have one or more complication
  - 47% have a visual acuity ≤ 1/10

Uveitis Subtypes in a German Interdisciplinary Uveitis Center — Analysis of 1916 Patients

EVA JAKOB, MIRJAM S. REULAND, FRIEDERIKE MACKENSEN, NADINE HARSCH, MONIKA FLECKENSTEIN, HANNS-MARTIN LORENZ, REGINA MAX, and MATTHIAS D. BECKER

J Rheumatol 2009

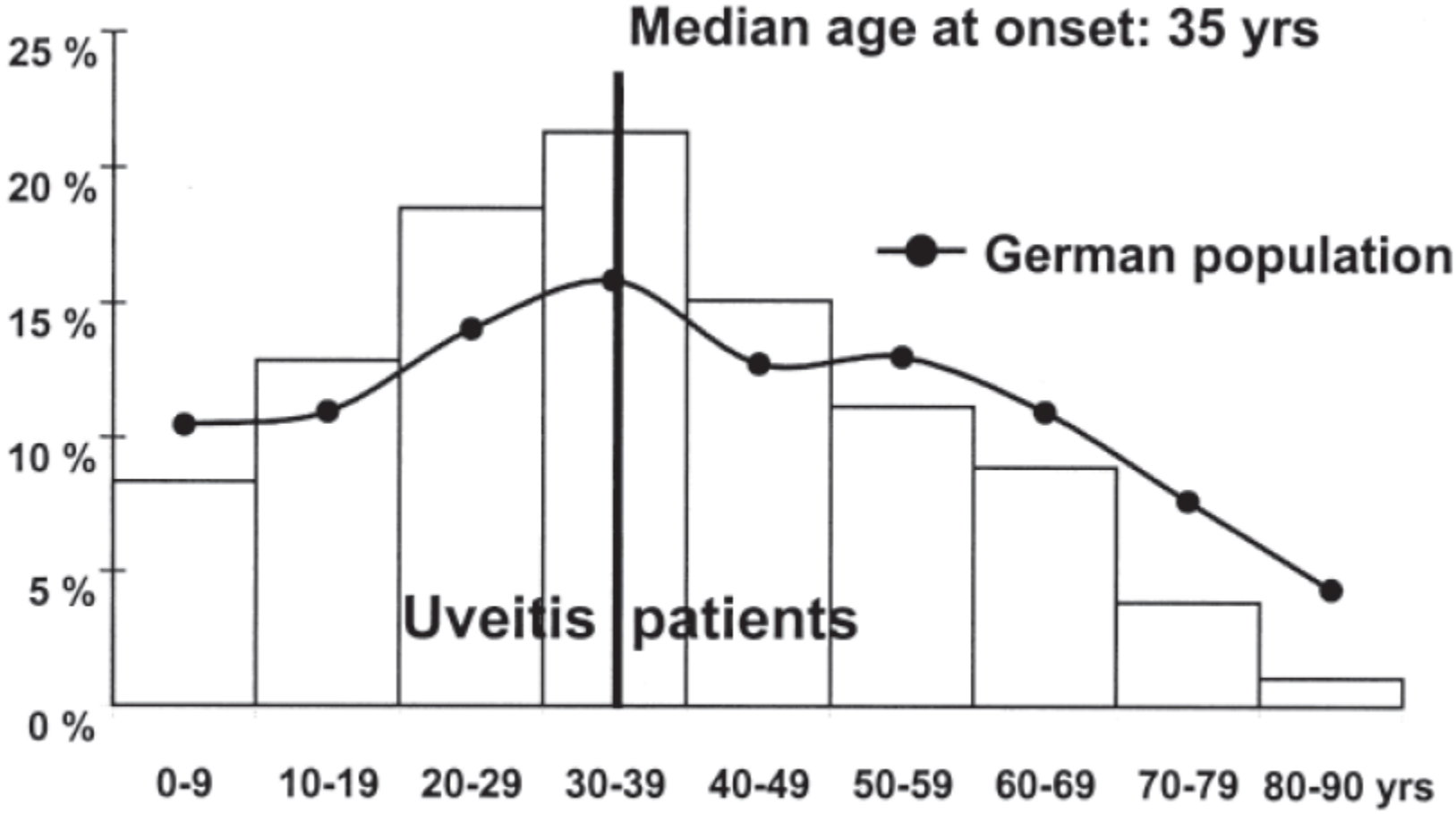


Figure 1. Age distribution at onset of uveitis manifestations in comparison to the German population pyramid (source: Statistisches Bundesamt[47]).

Incidence des uvéites selon les séries

Brézin Les uvéites
Ed Masson

	Nombre de patients	Référence	Centre	Uvéite antérieure	Uvéite intermédiaire	Uvéite postérieure	Panuvéite
Uvéites pédiatriques	251	Kimura, 1954 [3]	San Francisco	31,5 %	7,5 %	49 %	2 %
	130	Tutgal-Tutcun, 1996 [5]	Boston	58,4 %	20 %	13,8 %	7,6 %
	267	Pivetti-Pezzi, 1996 [6]	Rome	33,3 %	25,1 %	26,6 %	15 %
	219	Kadayifcilar, 2003 [7]	Ankara	43,38 %	11,87 %	31,05 %	13,7 %
	123	De Boer, 2003 [8]	Utrecht	36 %	24 %	19 %	21 %
	148	Rosenberg, 2004 [9]	Miami	30,4 %	27,7 %	23,7 %	18,2 %
	269	Kump, 2005 [10]	Boston	56,88 %	20,82 %	6,32 %	15,99 %
	276	BenEzra, 2005 [11]	Jerusalem	13,4 %	41,7 %	14,1 %	30,8 %
	163	Hamade, 2009 [12]	Riyadh	42 %	20 %	12 %	31 %
Uvéites de l'adulte	600	Henderly, 1987 [13]	Californie du Sud	28 %	15 %	38 %	18 %
	865	Rothova, 1992 [14]	Amsterdam	55 %	9 %	17 %	20 %
	558	Tran, 1994 [15]	Lausanne	61 %	10 %	21 %	7 %
	1 122	Paivonsalo-Hietanen, 1994 [16]	Turku	92 %	1 %	6 %	1 %
	1 237	Rodriguez, 1996 [17]	Boston	51,6 %	13 %	19,4 %	16 %
	1 417	Pivetti-Pezzi, 1996 [18]	Rome	49 %	12 %	22 %	16 %
	1 273	Biswas, 1996 [19]	Madras	39 %	17 %	29 %	15 %
	655	Merkanti, 2001 [20]	Verone	58 %	3 %	26 %	13 %
	1 233	Singh, 2004 [21]	Chandigarh	49,2 %	16,1 %	20,2 %	14,7 %
	1 752	Yang, 2005 [22]	Guangzou	45,6 %	6,1 %	6,8 %	41,5 %
	1 916	Jakob, 2008 [23]	Heidelberg	45,4 %	22,9 %	13,5 %	6,2 %
	488	Hamade, 2009 [12]	Riyadh	60 %	6 %	24 %	11 %

Uveitis in children

- Medical history, signs and symptoms
- Always try to play with children they should not be afraid of your examination
- In very young children if examination not possible do not hezitate to ask for a general anesthesia.
- Amblyopia should always be treated simultaneousely
- Helpful tool laser flare photometry, OCT
- Oral angiography possible in very young children

Aetiology according to the age of patients

Ocular Immunology & Inflammation, Early Online, 1–14, 2013

TABLE 1. Differential diagnosis of common causes of intermediate and posterior pediatric uveitis by age at presentation.

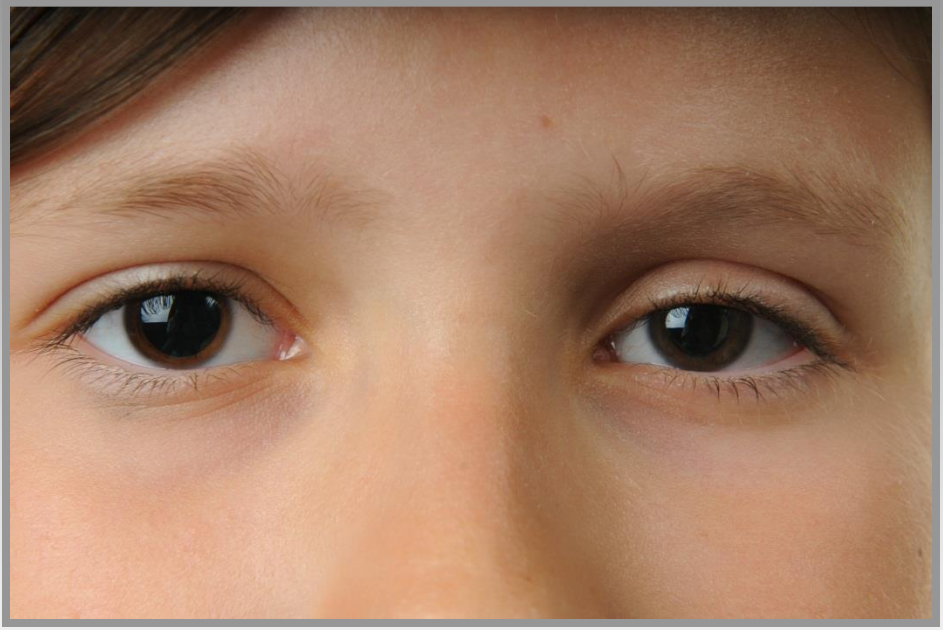
Infants (age 0–2 years)
- Infectious causes:
  - Toxoplasmosis
  - HSV retinitis
  - Toxocariasis
  - Rubella
  - Congenital syphilis
- Masquerade:
  - Retinoblastoma

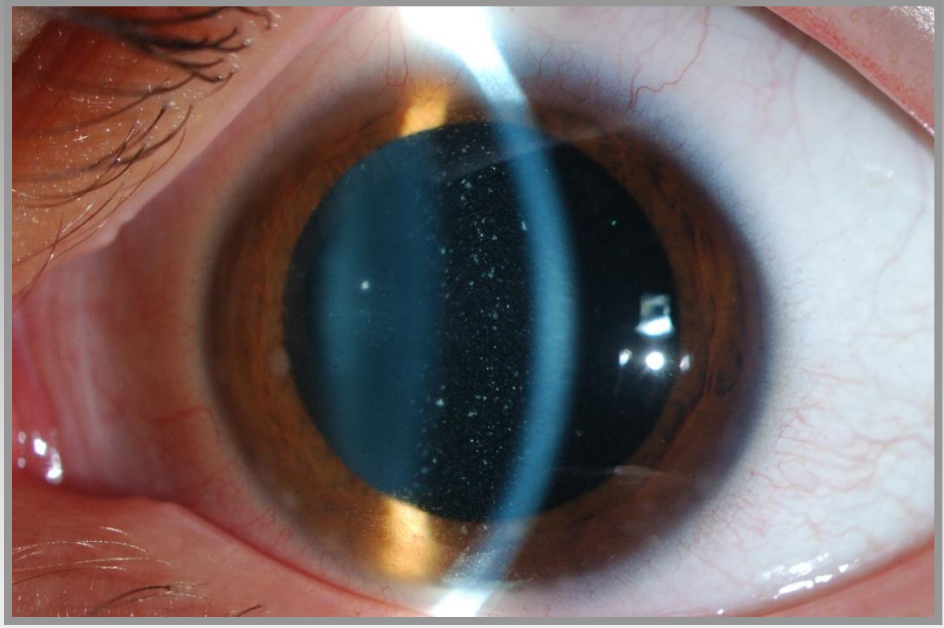
Children (2–10 years)
- Infectious:
  - Toxoplasmosis
  - Toxocariasis
  - Lyme disease
  - Cat-scratch disease
- Autoimmune:
  - Juvenile idiopathic arthritis
  - Familial juvenile systemic granulomatosis (Blau syndrome)
  - Chronic infantile neurological cutaneous and articular/ neonatal onset multisystem
- Masquerade:
  - Leukemia
  - Retinitis pigmentosa
  - Juvenile xanthogranuloma

Adolescents (10–20 years)
- Infectious:
  - Toxoplasmosis
  - Presumed ocular histoplasmosis syndrome
- Autoimmune:
  - Pars planitis
  - HLA-B27 associated disease
  - Sarcoidosis
  - Acute posterior multifocal placoid pigment epitheliopathy
  - Juvenile idiopathic arthritis
  - Fuchs heterochromic iridocyclitis
  - Tubulointerstitial nephritis and uveitis syndrome

Any age
- Infectious:
  - HIV Retinopathy
  - CMV retinitis
  - Acute retinal necrosis (VZ/HSV)
  - Endolphthalmitis
  - Lyme disease
  - Cat-scratch disease
  - Tuberculosis
- Autoimmune:
- Sarcoidosis
  - Tubulointerstitial nephritis and uveitis syndrome
  - Vogt-Koyanagi-Harada syndrome
  - Adamantiades-Behçet disease
- Masquerade:
  - Intraocular foreign body
  - Leukemia
  - Retinal detachment
  - Retinitis Pigmentosa
  - Juvenile Xanthogranuloma

Diffuse infiltrating retinobalstoma
Masquerade syndrome





E. M. 10 ans

Collaboration with prof F. Munier

Risk factor for ocular involvement in JIA

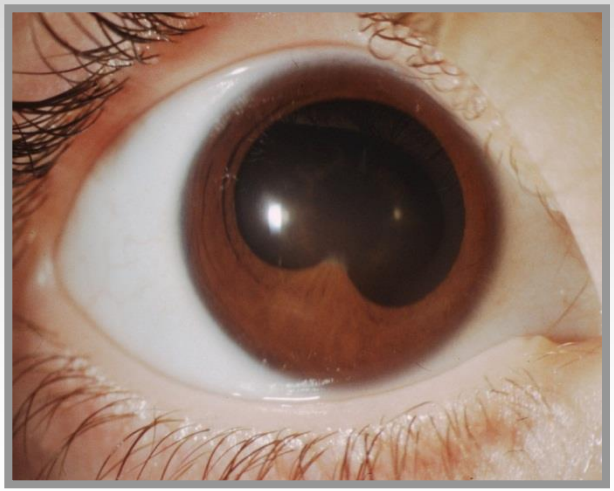
- Female (F:M ratio 3:1)
- Oligoarticular arthritis
- Young age at onset of arthritis
- ANA seropositivity 70-80% of cases
- RF seronegativity

Eular classification
7 classes

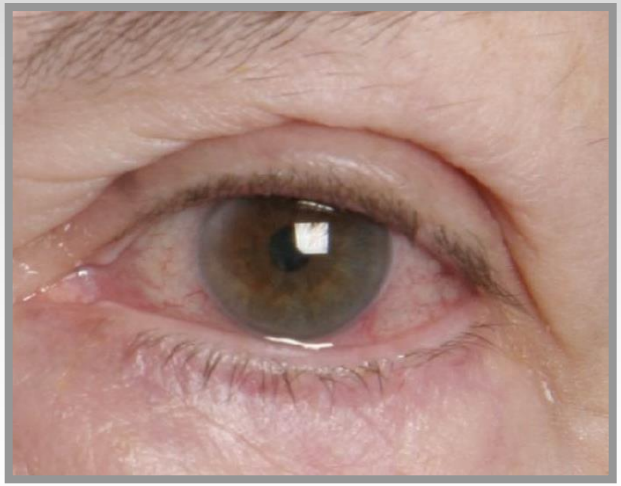
- Systemic onset JIA Uveitis extremely rare
- Oligoarticular JIA fewer than 5 joints involved during the first 6 months of the disease => most cases of chronic anterior uveitis
- Polyarticular JIA 10% with RF negative develop uveitis
- Polyarticular JIA 5 or more joints affected during the first 6 months, uveitis is rare in RF positive,
- Psoriatic arthritis Uncommon cause of JIA, uveitis present in 10% of cases
- Enthesitis related arthritis
- Undifferentiated arthritis

Uveitis in JIA

- Ocular inflammation in 2-34% of case
- Mostly bilateral (unilateral in the beginning)
- Anterior uveitis
- Mostly asymptomatic

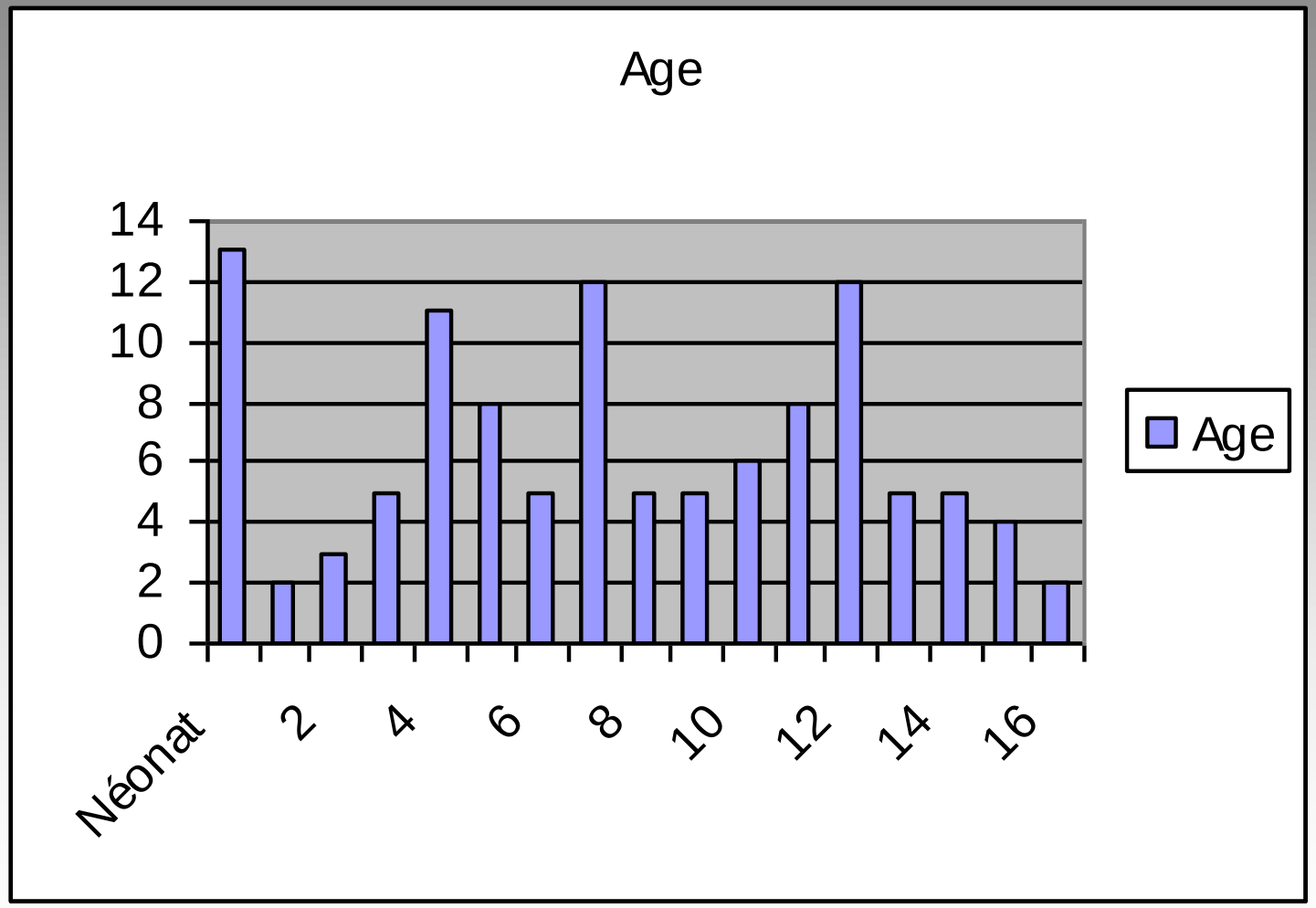


UAA in JIA

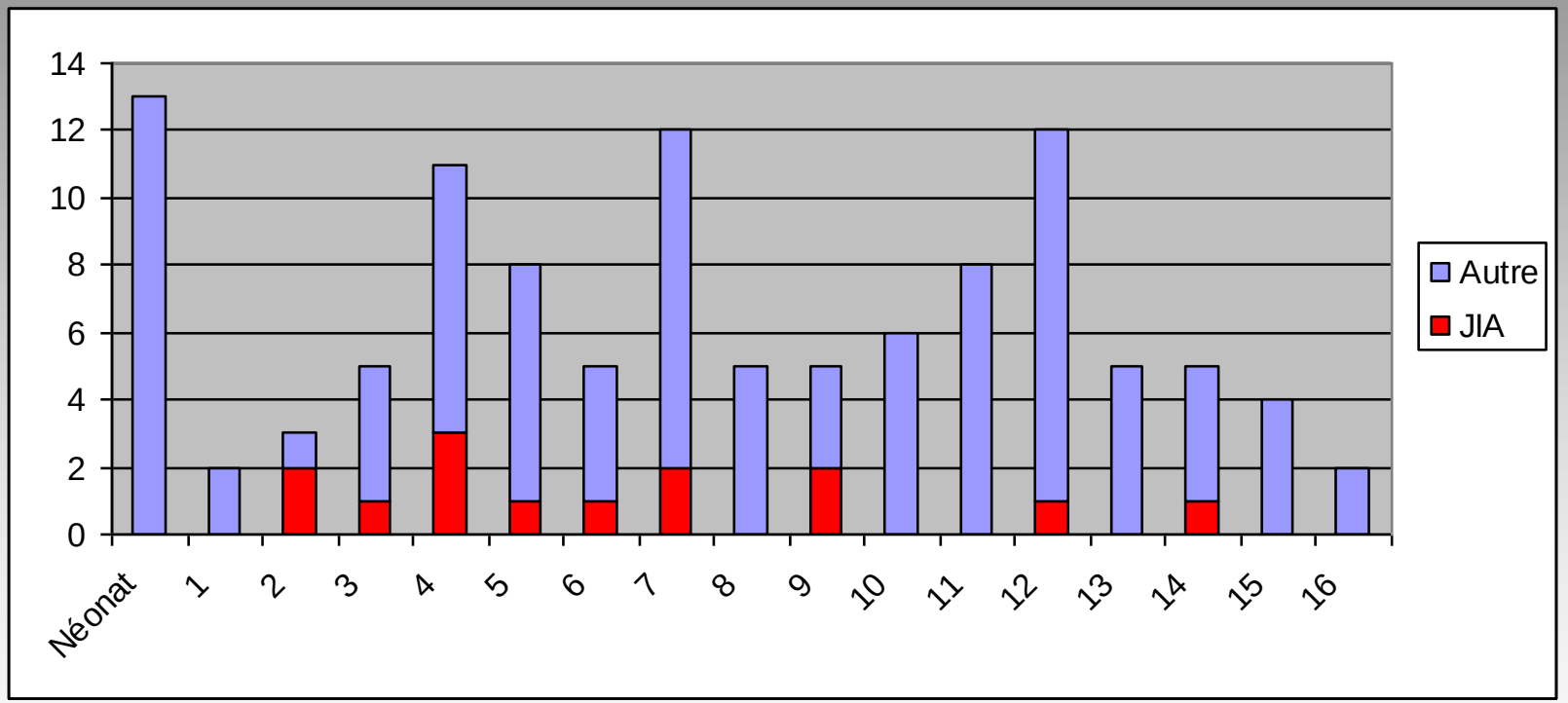


UAA HLA-B27

Distribution of patients age in paediatric uveitis between 1998 – 2008 111 new patients



Arthrite Juvénile Idiopathique (JIA)
14/111 in 1998 (13%)



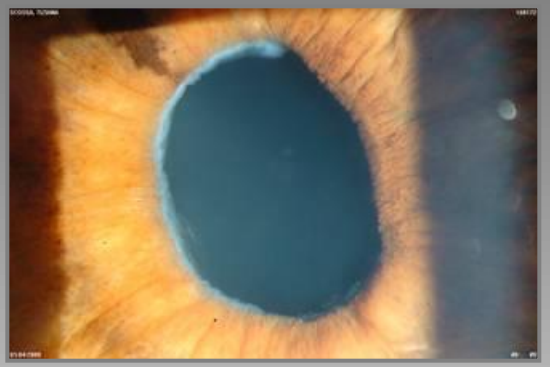
Complications in uveitis

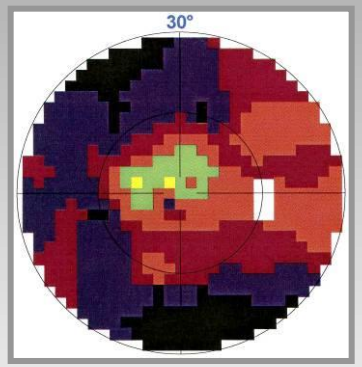
50% of blindness before the era of steroids

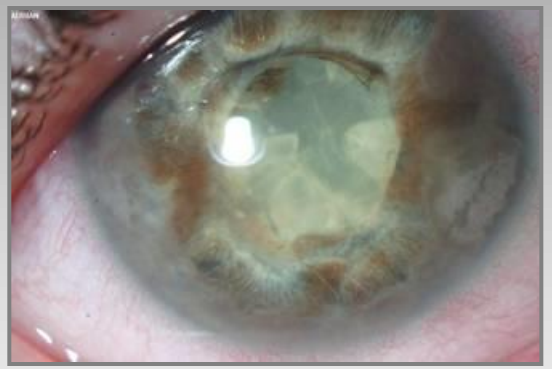
- Posterior synechiae
- Glaucoma
    - Inflammation
    - Steroid induced

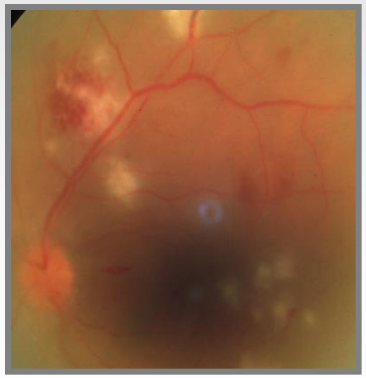
- Cataracte
- Band keratophathy

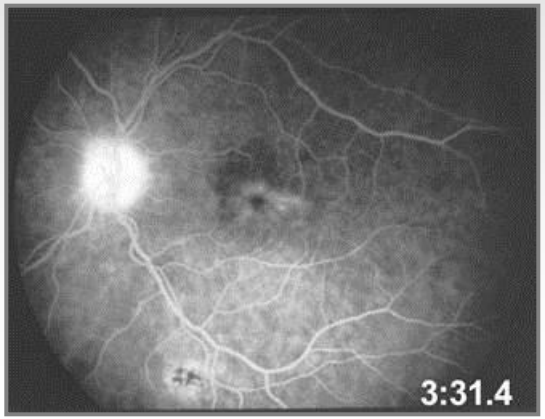
- Vitreous opacities
- Retinal lesions
    - Vasculitis => Vascular occlusions
    - Macular edema
    - Retinal necrosis
- Amblyopia





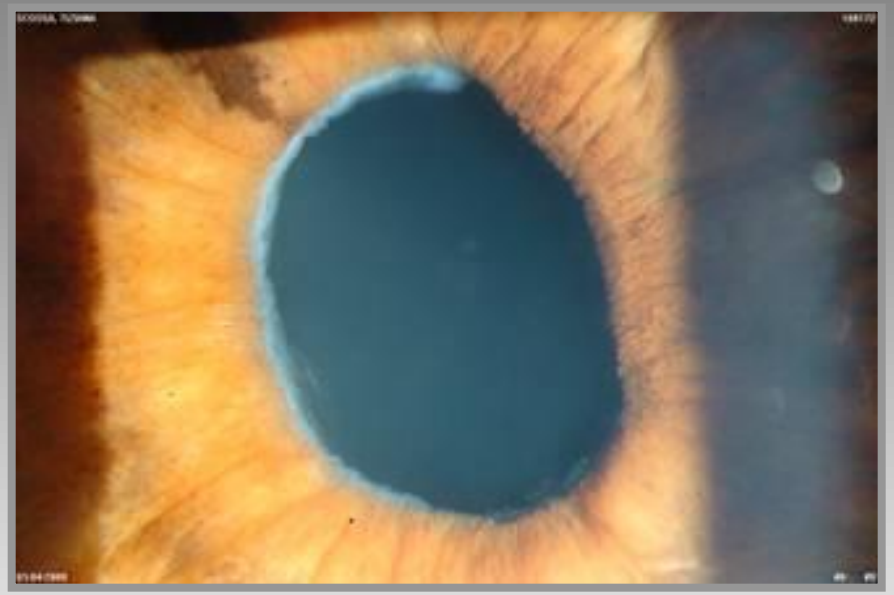


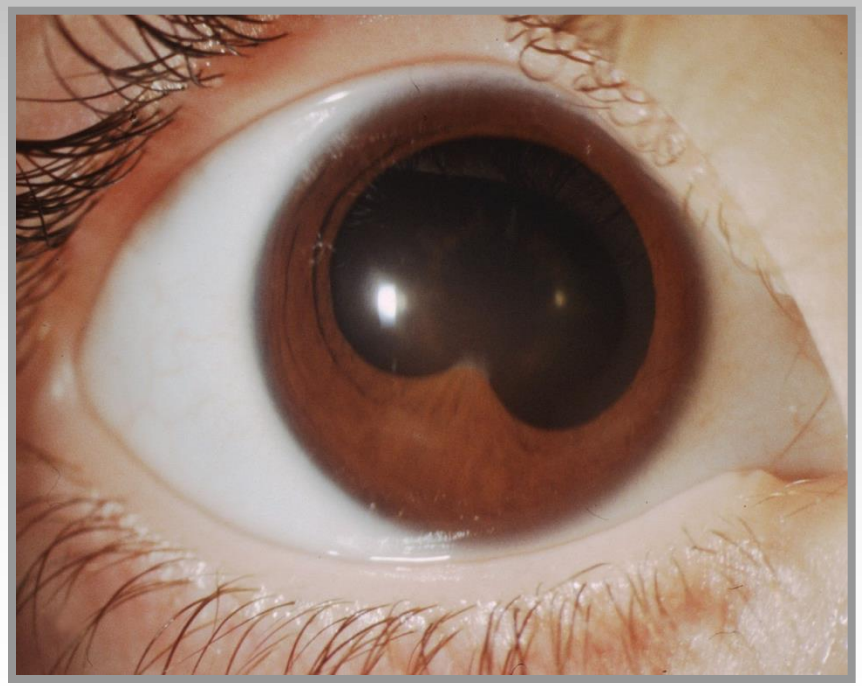


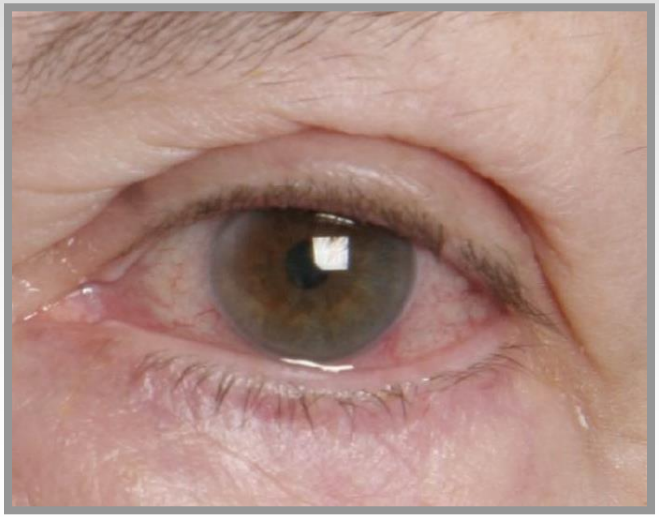


Complications des uvéites

1. Synéchies

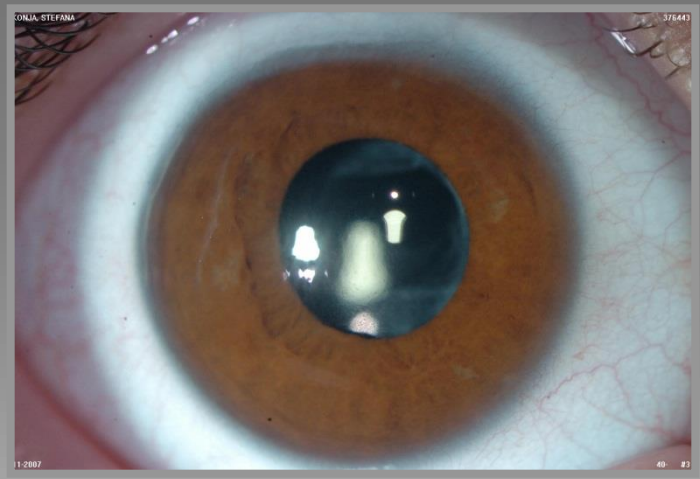


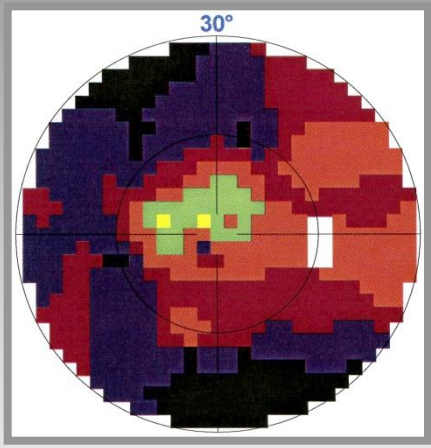


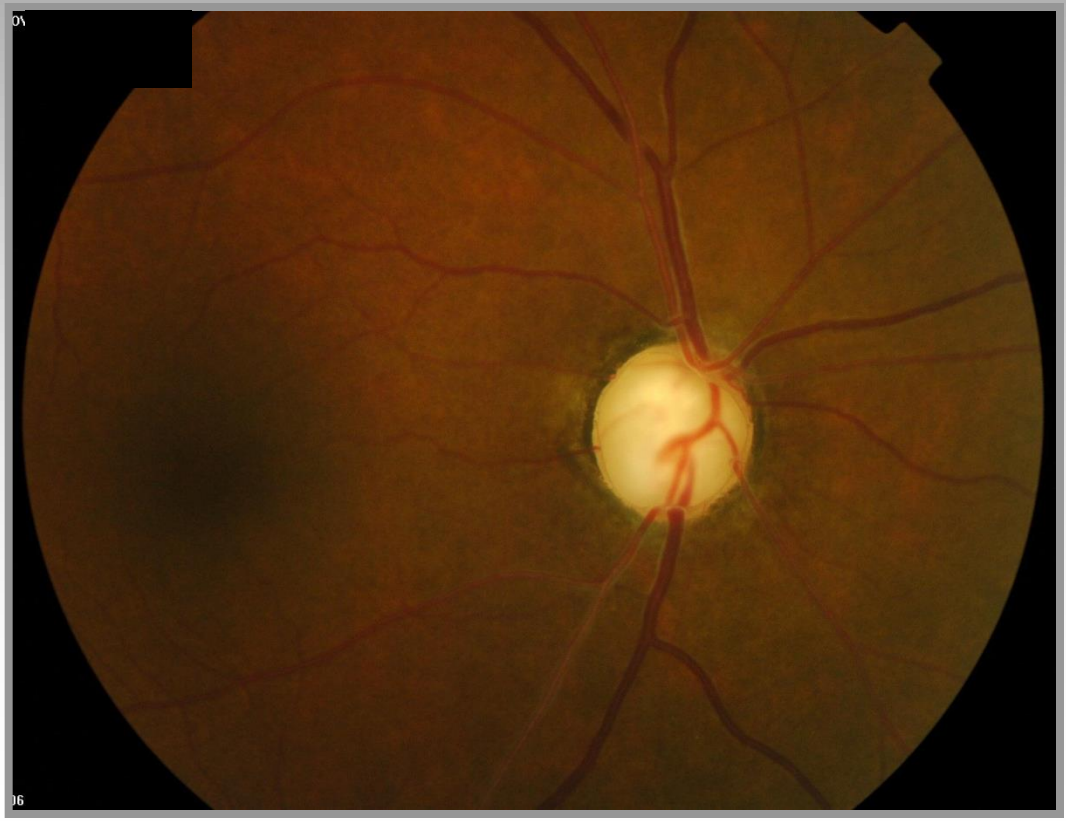


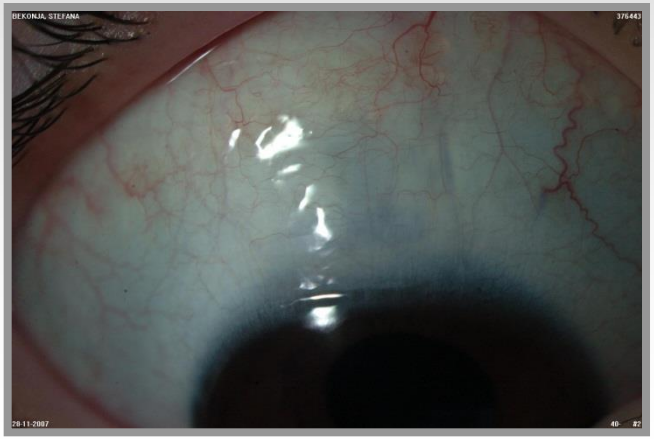
Complications

2. Glaucome







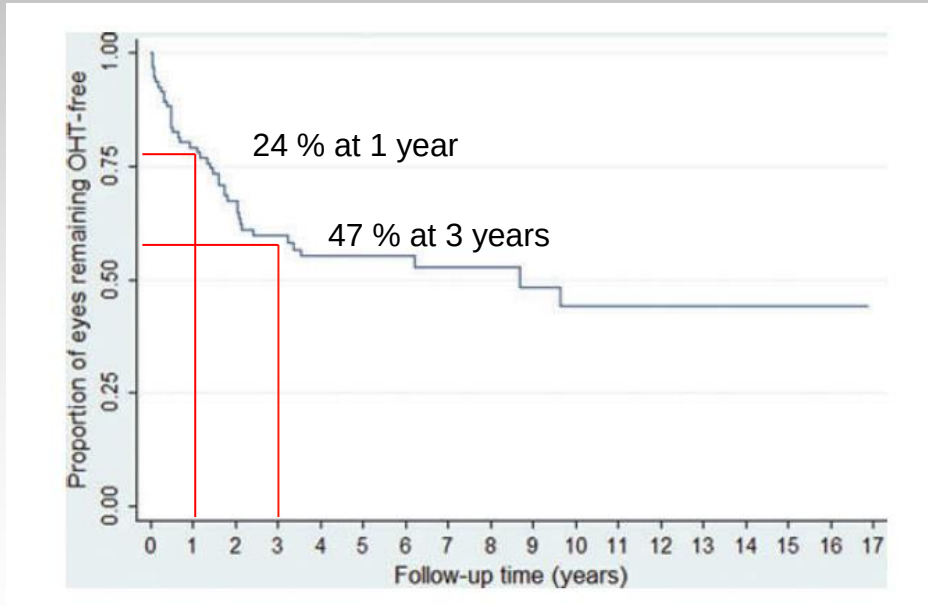


Occurrence of and Risk Factors for Ocular Hypertension and Secondary Glaucoma in Juvenile Idiopathic Arthritis-associated Uveitis

Inna G. Stroh MD, PhD, Ahmadreza Moradi MD, Bryn M. Burkholder MD, Dana M. Hornbeak MD, MPH, Theresa G. Leung MD & Jennifer E. Thorne MD, PhD

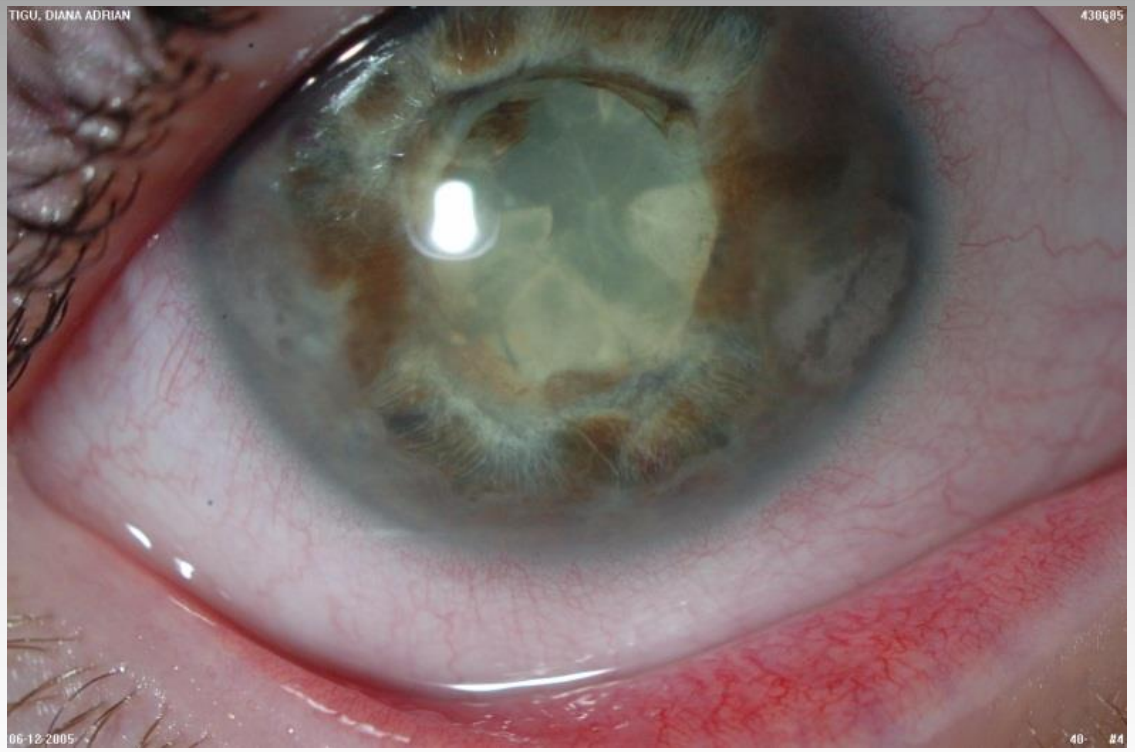
Ocular Immunology & Inflammation, 2016; 00(00): 1–10

- 24% at 1 year
- 47% at 3 years
- Risk factors for OHT
  - Ant uveitis aHR 8.2
  - Systemic corticosteroids at presentation aHR 5.34
- Protective factors for OHT
  - Early introduction of immuno-suppressive agents



Complications

3. Cataracte



Risk of Cataract Development among Children with Juvenile Idiopathic Arthritis-Related Uveitis Treated with Topical Corticosteroids

Jennifer E. Thorne, MD, PhD,[1,2] *Fasika A. Woreta, MD, MPH,*[1] *James P. Dunn, MD,*[1] *Douglas A. Jabs, MD, MBA*[2,3]

Ophthalmology 2010;117:1436–1441

- 0.01/EY if $\leq 3\ drops\ per\ day$
- 0.16/EY if $> 3 drops\ per\ day$

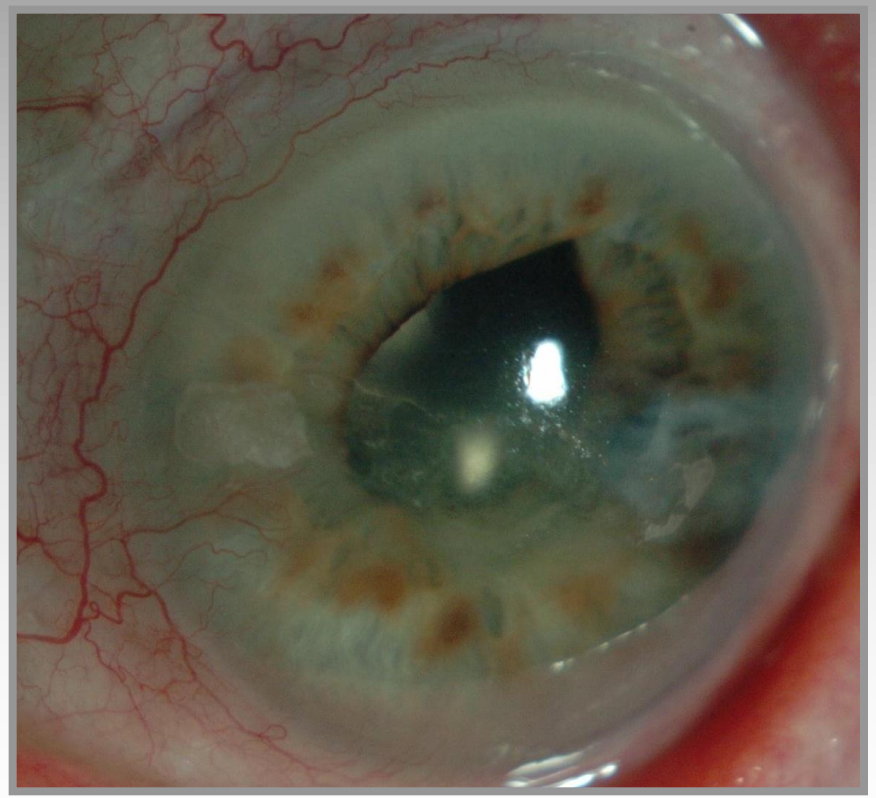
83% Less cataract if $\leq 3\ drops\ per\ day$

Table 3. Multivariate Time-Dependent Analysis of Use of Topical Corticosteroids and the Development of Cataract during Follow-up in Patients with Juvenile Idiopathic Arthritis-Related Uveitis

Characteristic	Relative Risk	95% Confidence Interval	*P* Value
Use of topical corticosteroids (≤3 drops daily vs. >3 drops daily)	0.13	0.02–0.69	0.02
Use of other forms of corticosteroids (yes vs no)	1.56	0.13–20.0	0.73
Use of immunosuppressive drug therapy (yes vs no)	0.96	0.31–2.94	0.94
Presence of active uveitis (yes vs no)	6.44	1.19–35.0	0.03

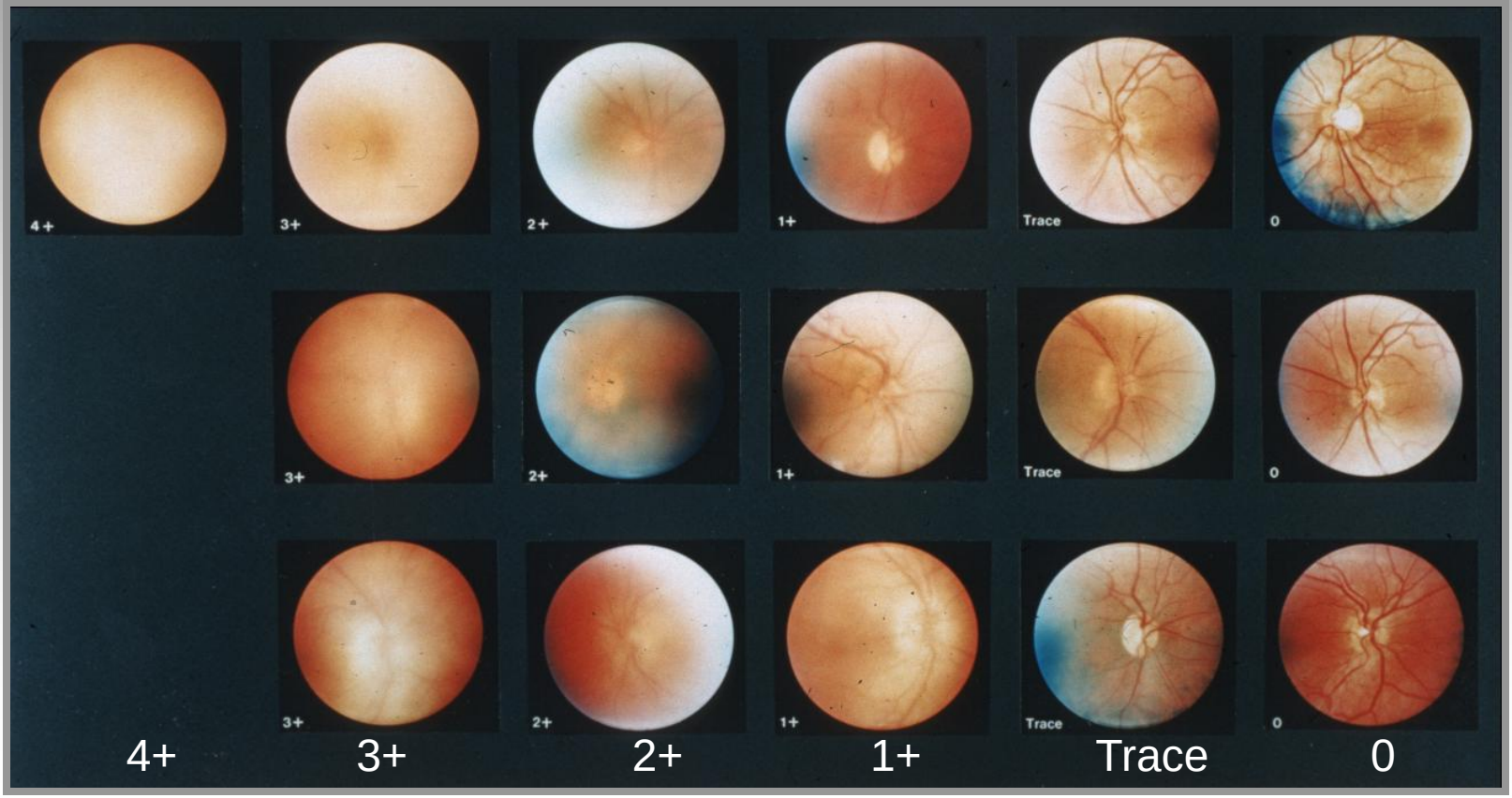
Complications

4. Kératite en bandelette



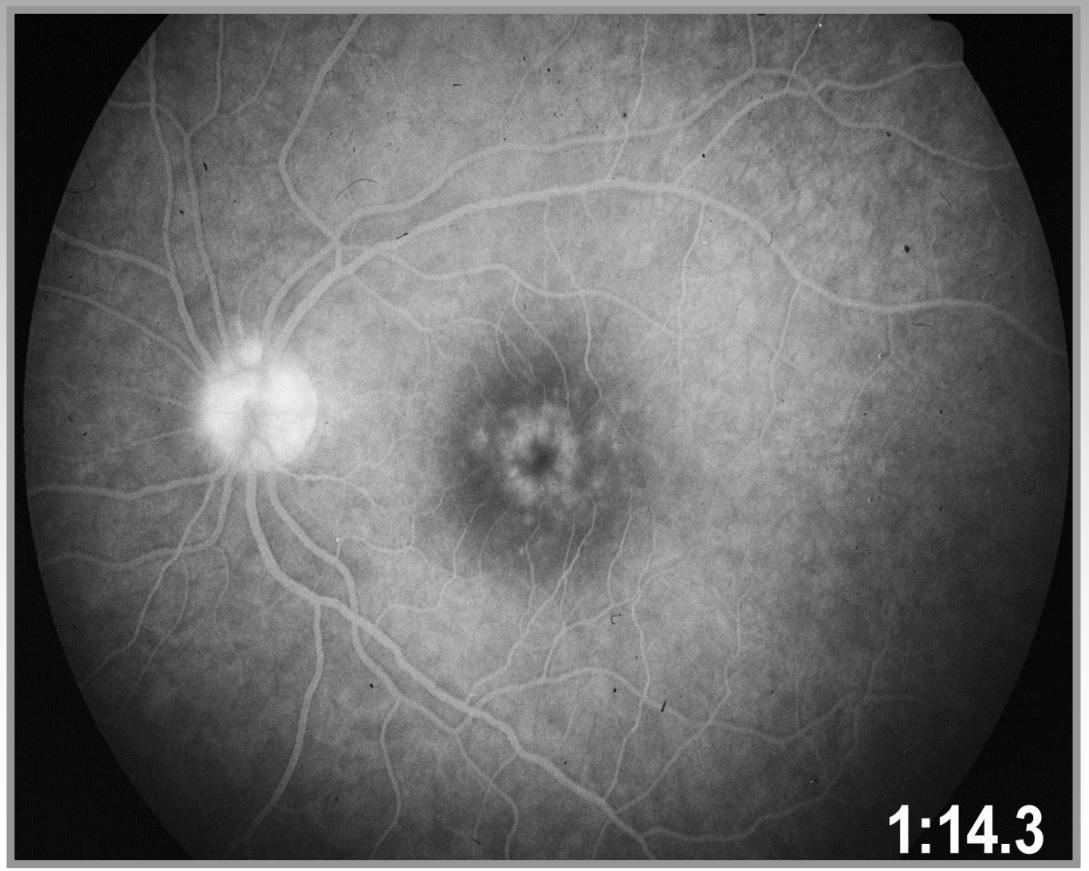
Complications

5. Vitrite sévère



Complications

5. Œdème maculaire



Complications

5. Amblyopie

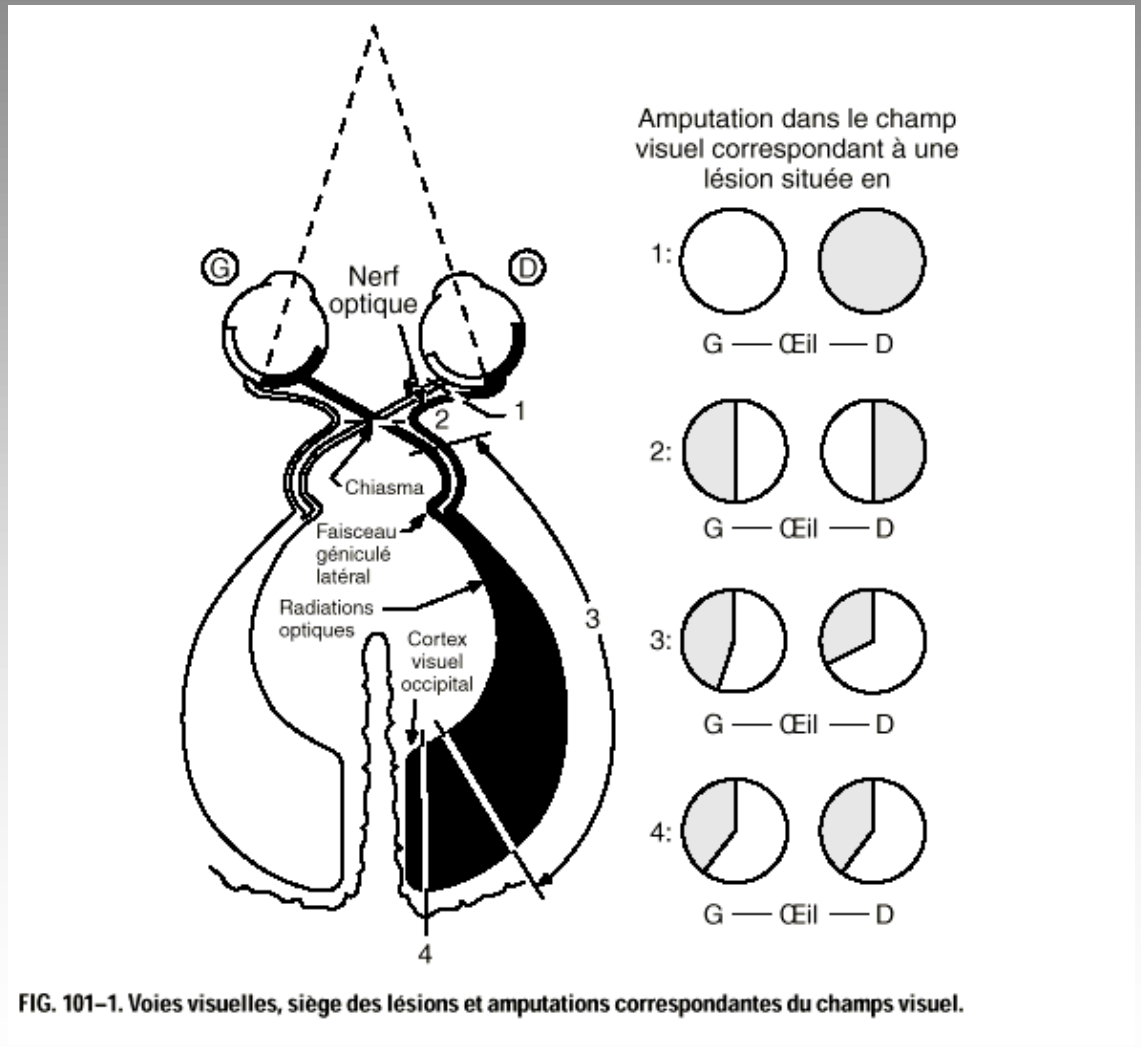


FIG. 101–1. Voies visuelles, siège des lésions et amputations correspondantes du champs visuel.

Risk Factors for Loss of Visual Acuity among Patients with Uveitis Associated with Juvenile Idiopathic Arthritis: The Systemic Immunosuppressive Therapy for Eye Diseases Study

Anthony C. Gregory II, MD, MPH,[1] John H. Kempen, MD, PhD,[2,3] Ebenezer Daniel, MBBS, MPH, PhD,[2] R. Oktay Kaçmaz, MD, MPH,[4,5] C. Stephen Foster, MD,[5,6] Douglas A. Jabs, MD, MBA,[7,8,9] Grace A. Levy-Clarke, MD,[10] Robert B. Nussenblatt, MD, MPH,[10] James T. Rosenbaum, MD,[11,12] Eric B. Suhler, MD, MPH,[11,13] Jennifer E. Thorne, MD, PhD,[1,7] for the Systemic Immunosuppressive Therapy for Eye Diseases Cohort Study Research Group

Ophthalmology 2013;120:186–192

- Population : 327 patients (596 affected eyes)
- Incidence rate for structural Ocular complication
  - Visual loss ≤ 20 / 50 at presentation 43%
  - Incidence of visual loss to the ≤ 20 / 50 = 0.18 Eye/year
  - Incidence of visual loss to the ≤ 20 / 200 = 0.09 Eye/year

  - Risk of ocular complication : 0.04 Eye / year (4 eyes/100p/1 year)

Complications during the follow-up median follow-up

Table 2. Incidence of Structural Ocular Complications and Loss of Visual Acuity in Juvenile Idiopathic Arthritis–Associated Uveitis among Those at Risk

Event	n/N*	Rate/PY†	95% CI	n/N‡	Rate/EY§	95% CI
Posterior synechiae	69/232	0.12	0.10–0.15	97/423	0.10	0.08–0.12
Band keratopathy	79/220	0.14	0.12–0.18	126/393	0.14	0.12–0.17
Macular edema	38/309	0.06	0.04–0.08	50/564	0.04	0.03–0.05
Epiretinal membrane	34/313	0.05	0.04–0.07	46/567	0.03	0.02–0.04
Hypotony	44/314	0.07	0.05–0.09	60/569	0.05	0.04–0.06
Ocular hypertension ≥21	86/281	0.17	0.15–0.22	125/508	0.10	0.08–0.11
Ocular hypertension ≥30	49/316	0.08	0.06–0.10	61/577	0.04	0.03–0.05
Any complication among naive¶	15/133	0.05	0.03–0.08	17/238	0.04	0.02–0.06
Any new complication#	133/327	0.17	0.15–0.20	221/596	0.15	0.13–0.17
VA ≤20/50**	91/209	0.20	0.16–0.24	131/356	0.18	0.16–0.21
VA ≤20/200**	70/257	0.14	0.11–0.17	95/452	0.09	0.08–0.11

Table 3. Risk Factors for Loss of Visual Acuity in Eyes with Juvenile Idiopathic Arthritis–Associated Uveitis

	≤20/50* (Crude†)			≤20/50* (Adjusted‡)			≤20/200* (Crude†)			≤20/200* (Adjusted‡)		
	HR	*95% CI*	*P Value*	*HR*	*95% CI*	*P Value*	*HR*	*95% CI*	*P Value*	*HR*	*95% CI*	*P Value*
Age at diagnosis (per year)	0.99	0.99–1.00	<0.01				0.99	0.99–1.00	<0.01			
Female gender	1.03	0.80–1.34	0.82				1.04	0.75–1.44	0.81			
Nonwhite race	1.33	1.02–1.73	0.03				2.63	1.99–3.46	<0.01			
Bilateral disease	0.89	0.54–1.44	0.63				0.59	0.35–1.01	0.05			
Uveitis duration	0.98	0.97–0.99	<0.01				0.99	0.98–1.01	0.34			
Posterior synechiae	1.63	1.30–2.05	0.01	1.58	1.25–1.99	<0.01	1.44	1.07–1.93	0.02	1.54	1.14–2.09	<0.01
Band keratopathy	1.17	0.94–1.44	0.13	1.20	0.97–1.49	0.10	1.38	1.06–1.81	0.02	1.32	1.00–1.74	0.05
Abnormal IOP§	1.26	0.99–1.61	0.06	1.33	1.03–1.70	0.03	1.14	0.83–1.56	0.42	1.10	0.79–1.51	0.59
Prior ocular surgery	1.76	1.39–2.25	<0.01	1.73	1.36–2.21	<0.01	3.40	2.35–4.92	<0.01	2.93	2.01–4.26	<0.01
AC cells‖												
0 AC cell grade	1.00			1.00				1.00			1.00	
0.5+ AC cell	1.37	1.12–1.69	<0.01	1.13	0.86–1.49	0.37	1.15	0.89–1.49	0.29	1.10	0.92–1.54	0.19
1+ AC cell	1.43	1.14–1.80	<0.01	1.41	1.12–1.78	<0.01	1.53	1.15–2.03	<0.01	1.37	1.03–1.84	0.03
2+ AC cell	1.51	1.13–2.03	<0.01	1.49	1.11–2.00	<0.01	1.80	1.29–2.53	<0.01	1.82	1.30–2.55	<0.01
3+ AC cell	1.74	1.11–2.73	0.02	1.68	1.07–2.64	0.02	3.33	2.18–5.08	<0.01	2.43	1.46–4.06	<0.01
4+ AC cell	2.45	0.91–6.62	0.08	2.23	0.83–6.01	0.11	6.99	3.29–14.84	<0.01	6.42	2.98–13.84	<0.01
Any vitreous cell or haze‖	1.29	1.05–1.59	0.02	1.32	1.07–1.62	0.01	1.43	1.10–1.95	<0.01	1.50	1.14–1.97	<0.01
Use of oral corticosteroids‖	1.26	1.02–1.56	0.03	1.43	1.15–1.79	<0.01	1.29	0.99–1.69	0.06	1.31	1.00–1.73	0.05
Use of IMT‖	0.39	0.21–0.74	<0.01	0.40	0.21–0.75	<0.01	0.79	0.57–1.08	0.14	0.80	0.62–1.02	0.08
logMAR score¶	1.09	0.94–1.27	0.25	1.13	0.97–1.32	0.12	2.47	2.13–2.87	<0.01	2.43	2.08–2.86	<0.01

AC = anterior chamber; CI = confidence interval; HR = hazard ratio; IMT = immunomodulatory therapy; logMAR = logarithm of the minimum angle of resolution.

*Characteristic was assessed as number of new events of vision loss per EY of follow-up. Vision was assessed using logMAR ($-\text{Log}_{10}$ Minimal Angle of Resolution) charts.

†Crude refers to univariate Cox regression analyses.

‡Adjusted refers to multivariate Cox regression analyses.

§An abnormal IOP was defined as >21 mmHg (ocular hypertension) or <5 mmHg (hypotony).

‖Time-updated analysis.

¶Per increase in 1 line of VA <20/15 at initial visit.

Analysis of Pediatric Uveitis Cases at a Tertiary Referral Center

Leila I. Kump, MD,[1,2] René A. Cervantes-Castañeda, MD,[1,2] Sofia N. Androudi, MD,[1,2] C. Stephen Foster, MD, FACS[1,2,3]

Ophthalmology 2005;112:1287–1292

Table 3. Diagnoses

Etiology	No. of Cases
Juvenile idiopathic arthritis–associated iritis	89
Pars planitis idiopathic	56
Anterior idiopathic uveitis	52
Panuveitis, idiopathic	22
Toxoplasmosis	9
Iritis, HLA-B27 associated	5
Acute retinal necrosis	3
Anterior uveitis tubulointerstitial nephritis	3
Posterior idiopathic uveitis	4
Multifocal choroiditis and panuveitis	3
Anterior uveitis, Kawasaki related	2
Idiopathic vasculitis	2
Panuveitis secondary to pseudotumor	2

Table 3. Diagnoses

Etiology	No. of Cases
Sarcoid uveitis	2
Sclerouveitis (posterior)	2
Vogt–Koyanagi–Harada syndrome	2
Adamantiades–Behçet disease	1
Herpes simplex virus keratouveitis	1
Idiopathic choroidal granuloma	1
Infectious endophthalmitis	1
Masquerade syndrome, late-onset retinoblastoma	1
Neuroretinitis, idiopathic	1
Sarcoid, suspected	1
Systemic lupus erythematosus	1
Sympathetic ophthalmia	1
Toxocariasis	1
Varicella-zoster virus iritis	1
Total	269

Ocular complications

Kump et al · Analysis of Pediatric Uveitis Cases at a Tertiary Referral Center

Table 4. Complications Encountered in 469 Eyes

Complication	Juvenile Idiopathic Arthritis (n = 165)		Anterior Idiopathic Uveitis (n = 98)		Intermediate Uveitis (n = 104)		Other (n = 102)	
	n	%	n	%	n	%	n	%
Cataract	105	64	28	29	32	31	23	22
Glaucoma	33	20	3	3	26	25	9	9
Band keratopathy	76	46	9	9	15	14	7	7
Posterior synechiae	96	58	25	26	24	23	19	19
Hypotony	17	10	0	0	1	1	0	0
Maculopathy (edema, cystic changes, scar)	43	26	3	3	41	39	24	24
Retinal detachment	5	3	1	1	3	3	5	5
Vitreous hemorrhage	0	0	0	0	3	3	2	2
Rubeosis	0	0	1	1	2	2	1	1
Epiretinal/neovascular membranes	17	10	1	1	19	18	15	15
Papilledema/papillitis	5	3	4	4	10	10	19	19

International consensus for therapy Heiligenhaus et al

- Topical corticosteroids
  < 3 drops/day for <3 months
- Oral prednisone 1mg/kg/j.
  Progressve tapering down.
- Immunosuppressive drug
  méthotrexate ou azathioprine
- Biologic (anti-TNFα)

Adapted from Evidence based interdisciplinary guidelines
Heiligenhaus et al, Rheumatol Int 2012: 32 1121-1133

TABLE 1. Frequency of Ophthalmologic Examination in Patients With Juvenile Idiopathic Arthritis Adapted From[4]

ANA Status/Pattern of Arthritis	Age at Diagnosis (yr)	Disease Duration (yr)
Ophthalmology exam every 3 mo		
ANA + oligoarticular	≤6	≤4
ANA + polyarticular	≤6	≤4
Ophthalmology exam every 6 mo		
ANA + oligoarticular	≤6	>4
ANA + polyarticular	≤6	>4
ANA + oligoarticular	>6	≤4
ANA + polyarticular	>6	≤4
ANA − oligoarticular	≤6	≤4
ANA − polyarticular	≤6	≤4
Ophthalmology exam every 12 mo		
ANA + oligoarticular	>6	>4
ANA + polyarticular	>6	>4
ANA + oligoarticular	≤6	>7
ANA + polyarticular	≤6	>7
ANA − oligoarticular	≤6	>4
ANA − polyarticular	≤6	>4
ANA − oligoarticular	>6	N/A
ANA − polyarticular	>6	N/A
Systemic	N/A	N/A

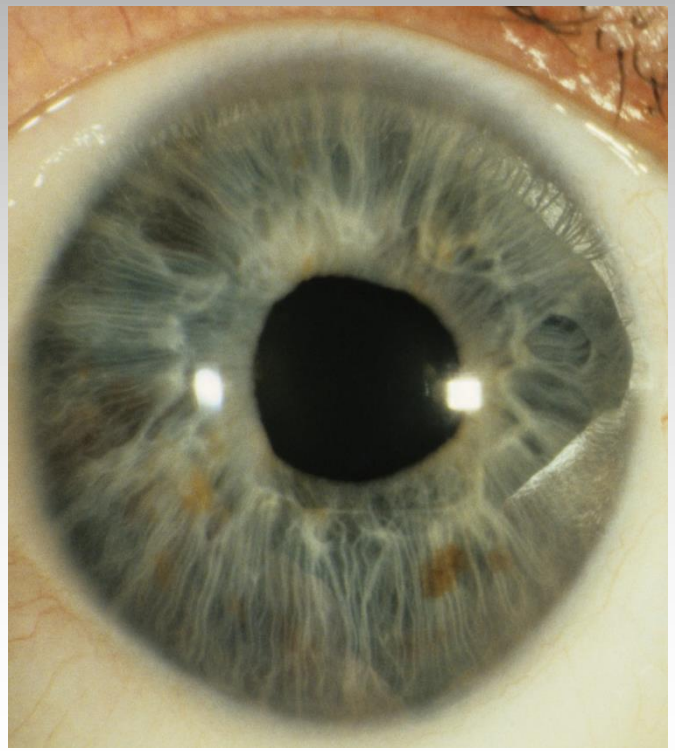
Tabelle 3
Empfehlungen der American Academy of Pediatrics, Sections of Rheumatology and Ophthalmology, bezüglich der Häufigkeit von ophthalmologischen Screening-Untersuchungen bei Kindern mit juveniler Arthritis (JIA) [5].

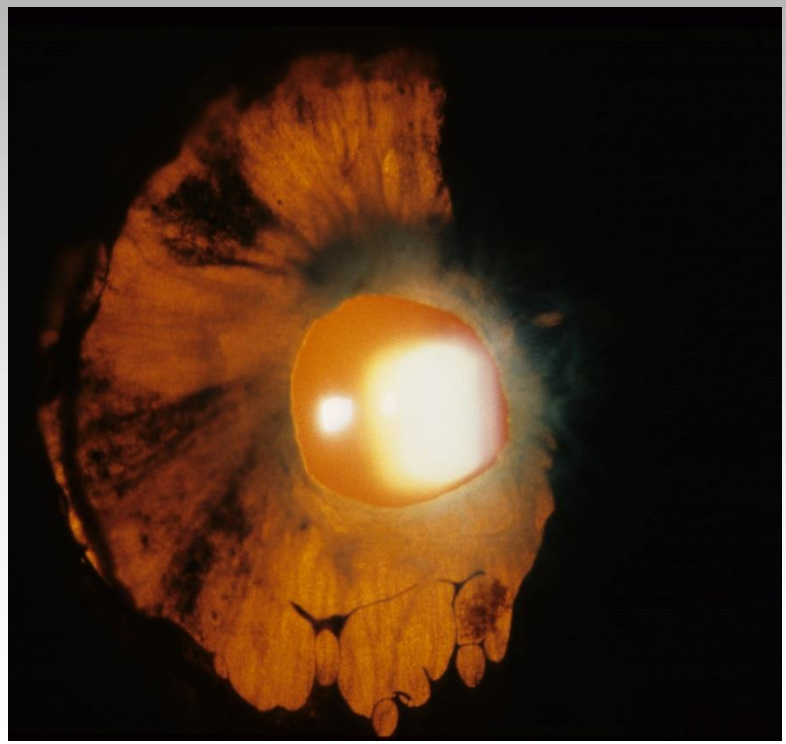
Alter bei Diagnose JIA	ANA-Status	Dauer der JIA	Screening-Häufigkeit
<6 Jahren	pos	<4 Jahren	3-monatlich
>6 Jahre	pos	<4 Jahren	6-monatlich
<6 Jahren	neg	<4 Jahren	6-monatlich
<6 Jahren	pos	4–7 Jahre	6-monatlich
>6 Jahre	pos	>4 Jahre	jährlich
>6 Jahre	neg	>4 Jahre	jährlich
<6 Jahren	pos	>7 Jahre	jährlich

Schweiz Med Forum 2012;12(38):739–741

Kératouvéite herpétique

- Patiente de 35 ans, uvéite antérieure unilatérale, granulomateuse, fond d'oeil normal



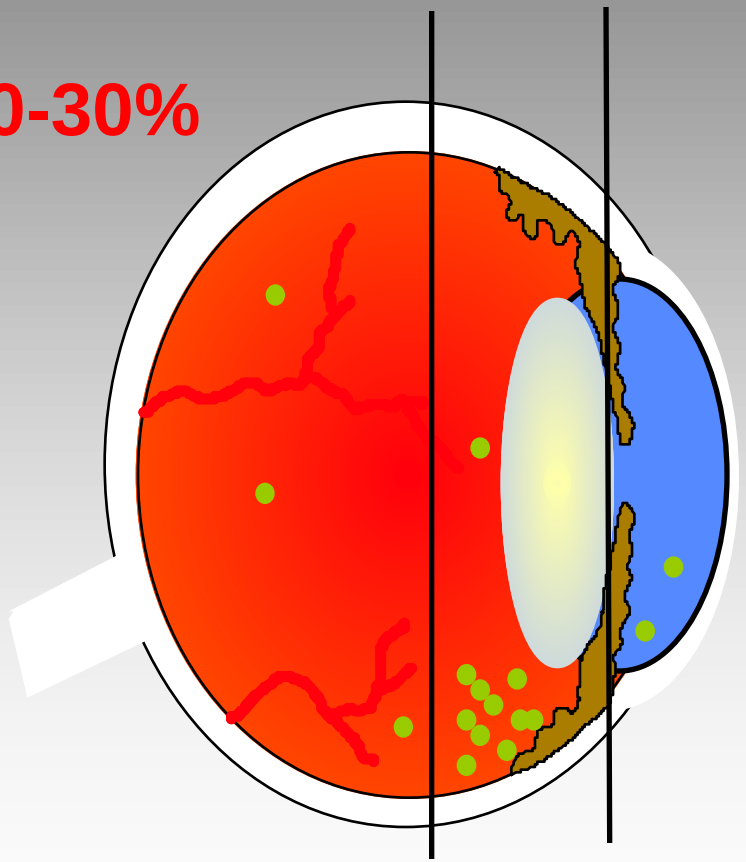


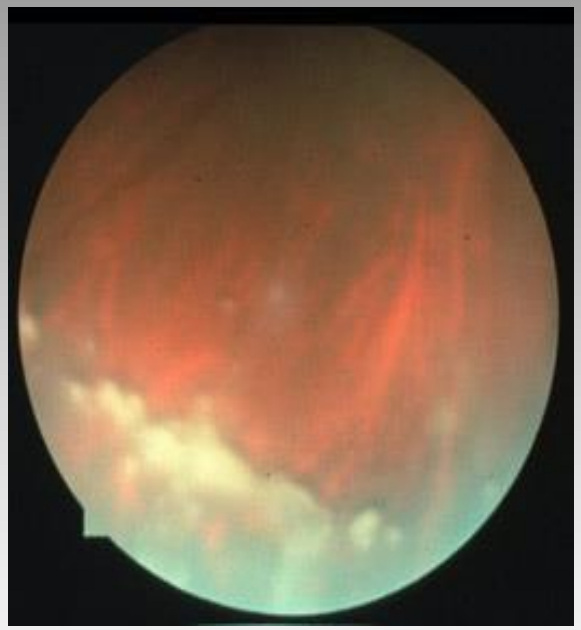
A_2. Anatomical classification

Intermediate uveitis 7%

Pars Planitis

Posterior 10-30%



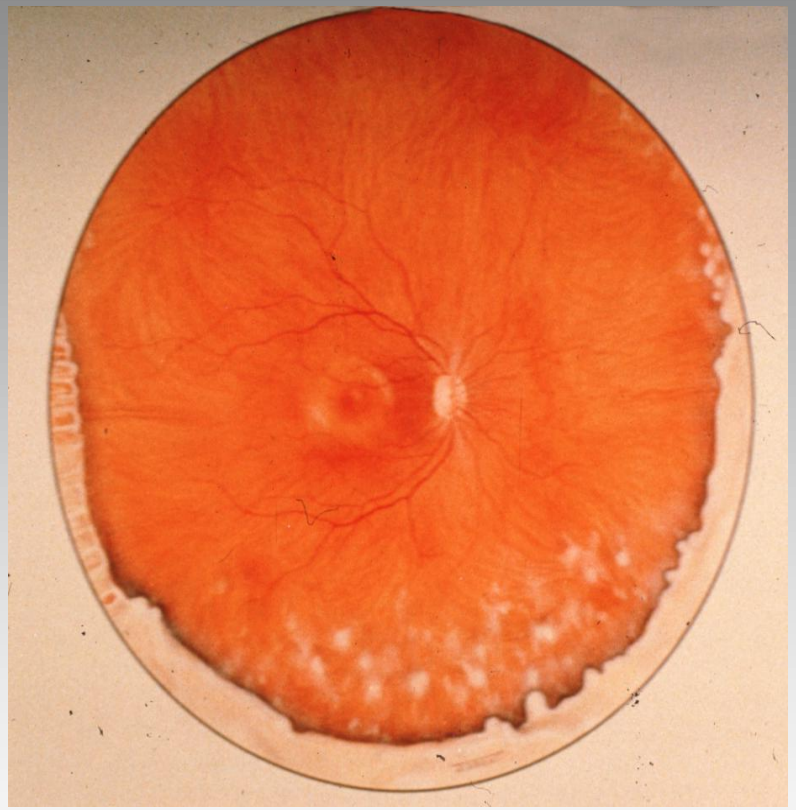


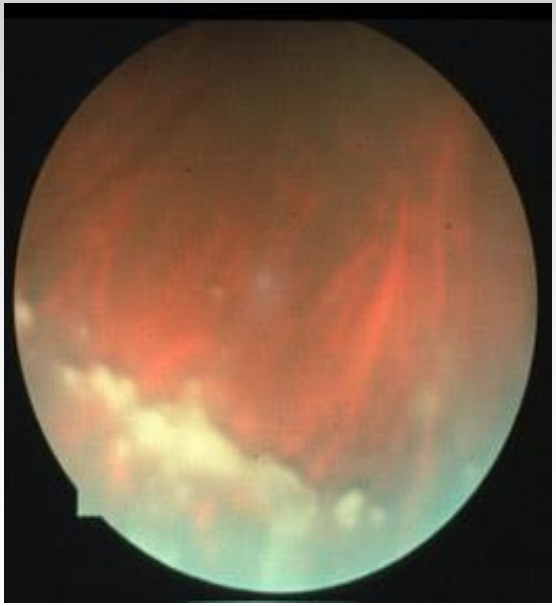
Anterior 50-60%

Panuveitis 7%

Intermediate uveitis

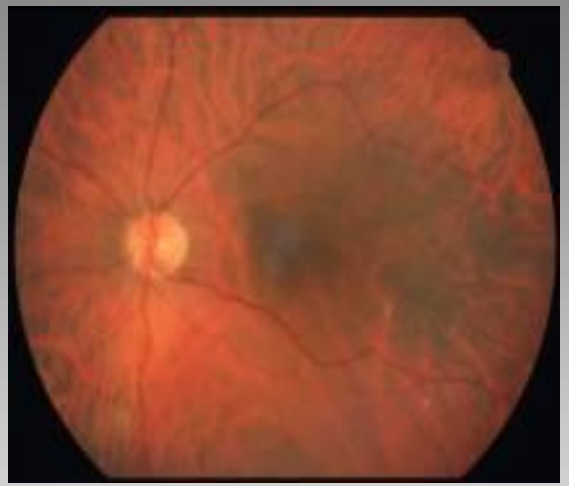
- Vitreous cells
- White exsudates «Snowballs»
- Snowbanking
- Macular Edema

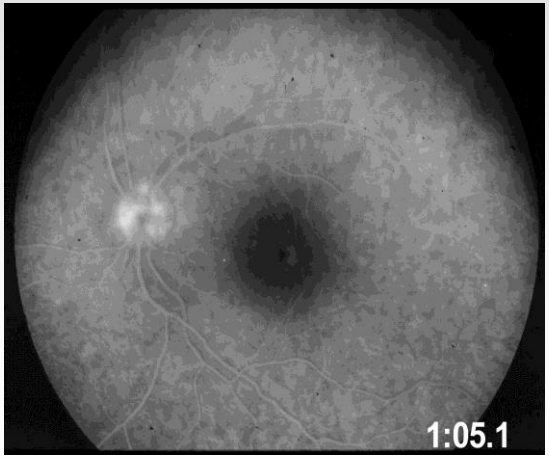


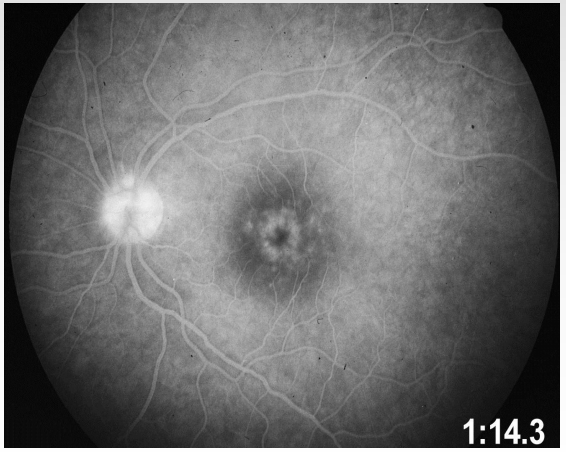


Macular edema

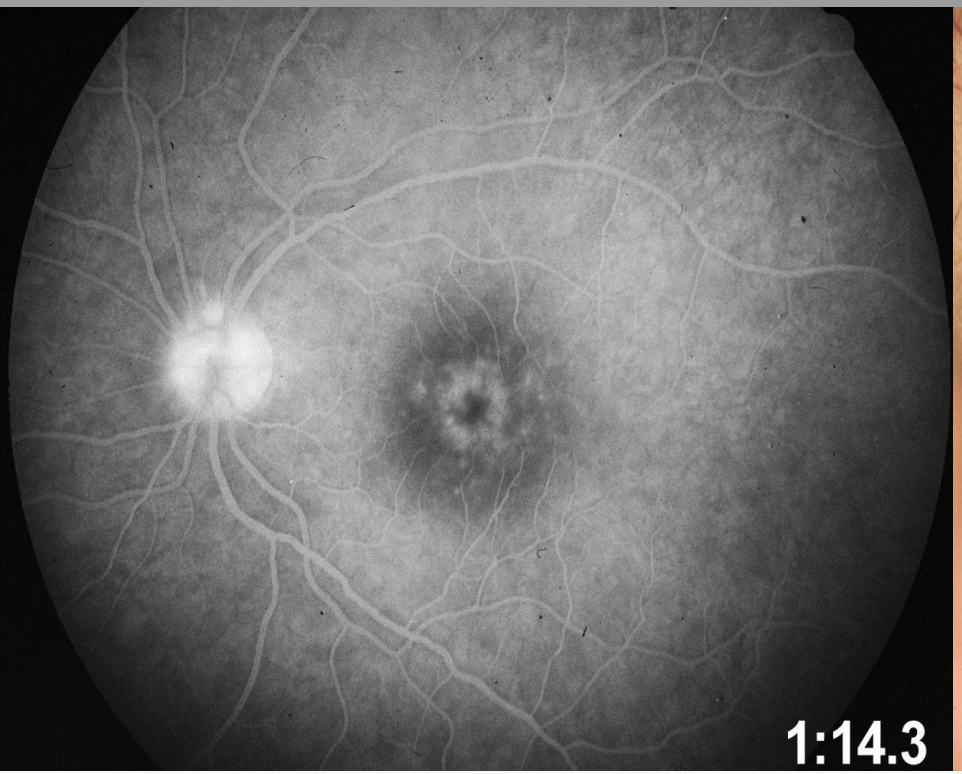
- No symptoms
  angiographic OCT
- Decrease in VA
- Photophobia
- Visual discomfort
- Blurred vision



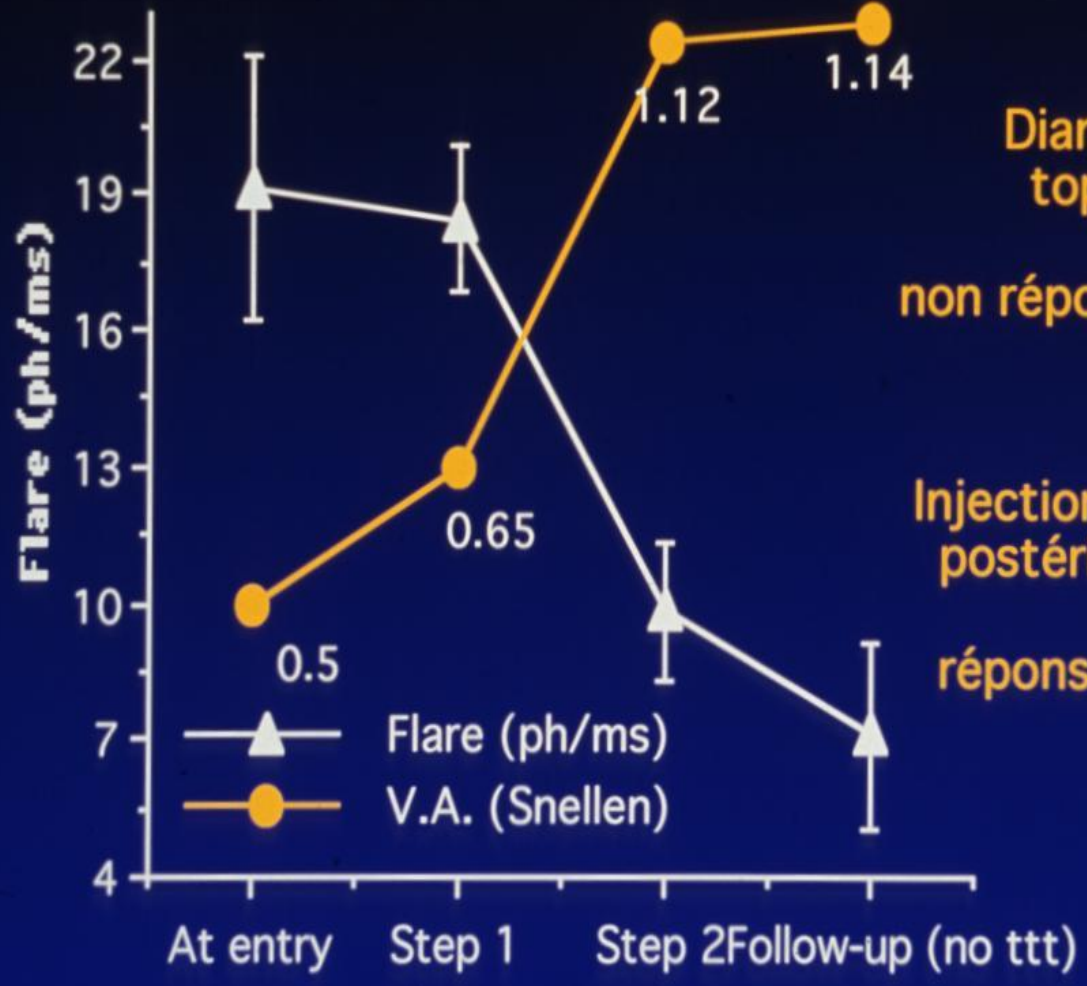




Posterior subtenon's steroid injection Kenacort 40 mg



Evolution du flare et de l'acuité visuelle dans les pars planites avec OMC (n = 10)



Etape 1

Diamox + traitement topique diclofenac

=>

non réponse 10 / 10 patients

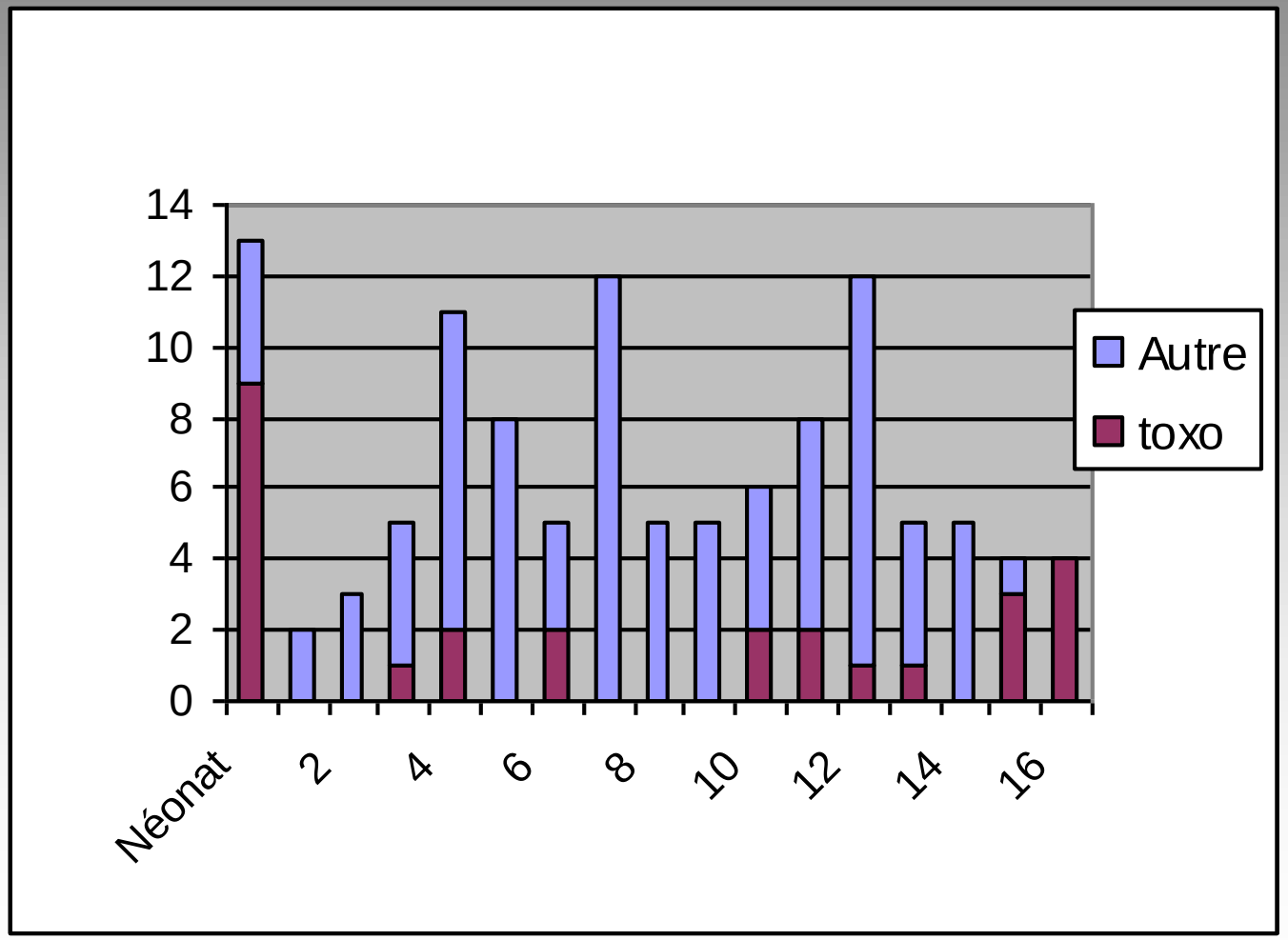
Etape 2

Injections sous-ténoniennes postérieures de stéroides

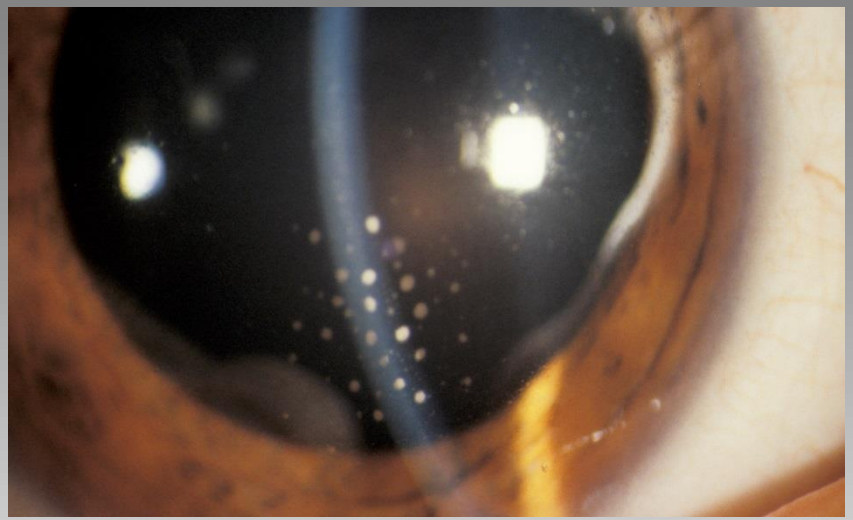
=>

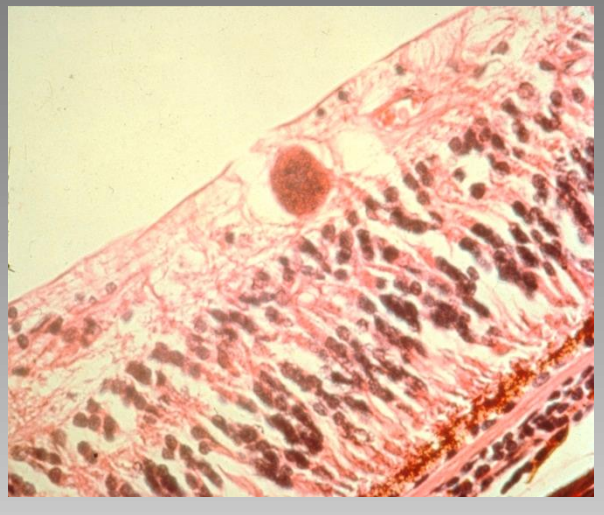
réponse 10 / 10 patients

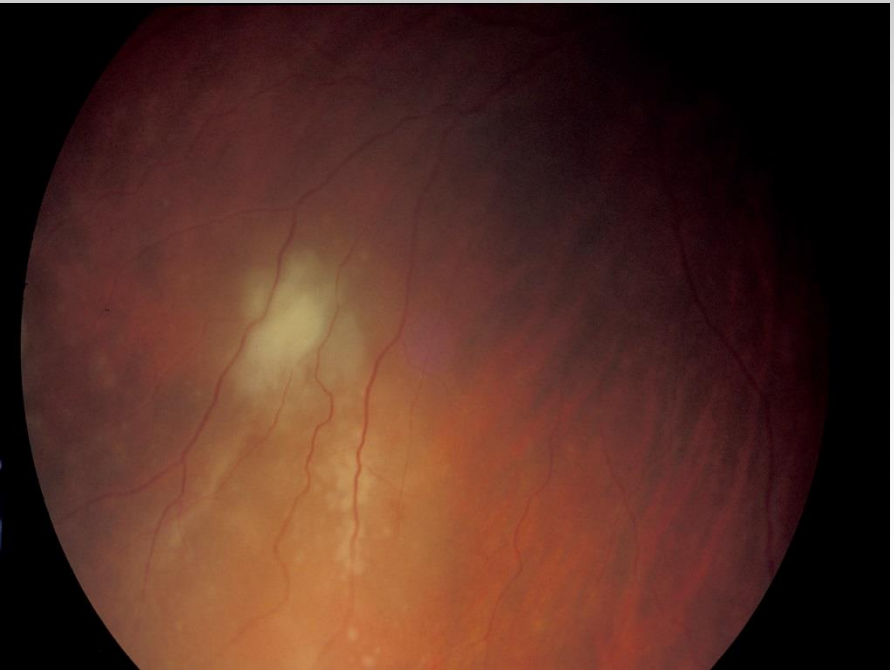
Toxoplasmoses enfant n = 27/111 (25%)

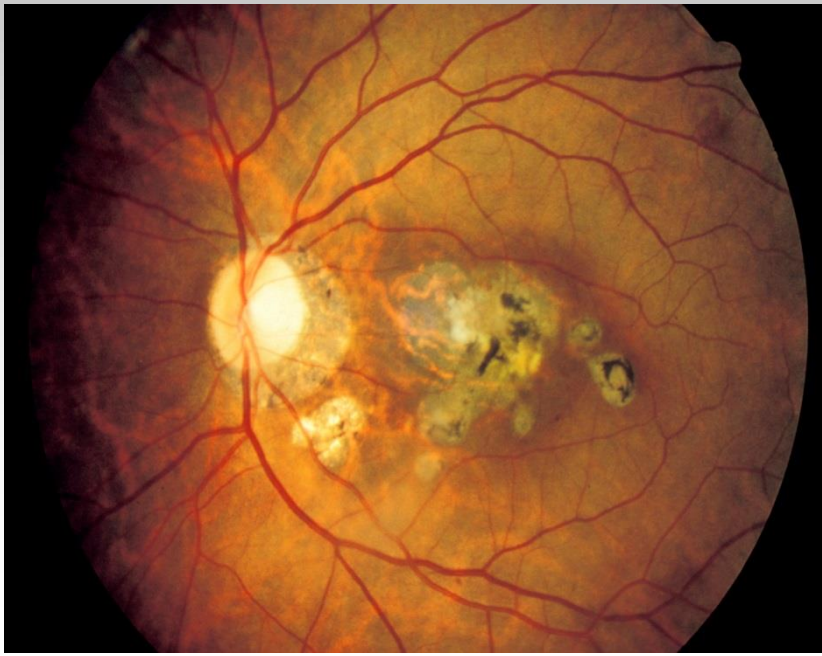


Ocular toxoplamosis







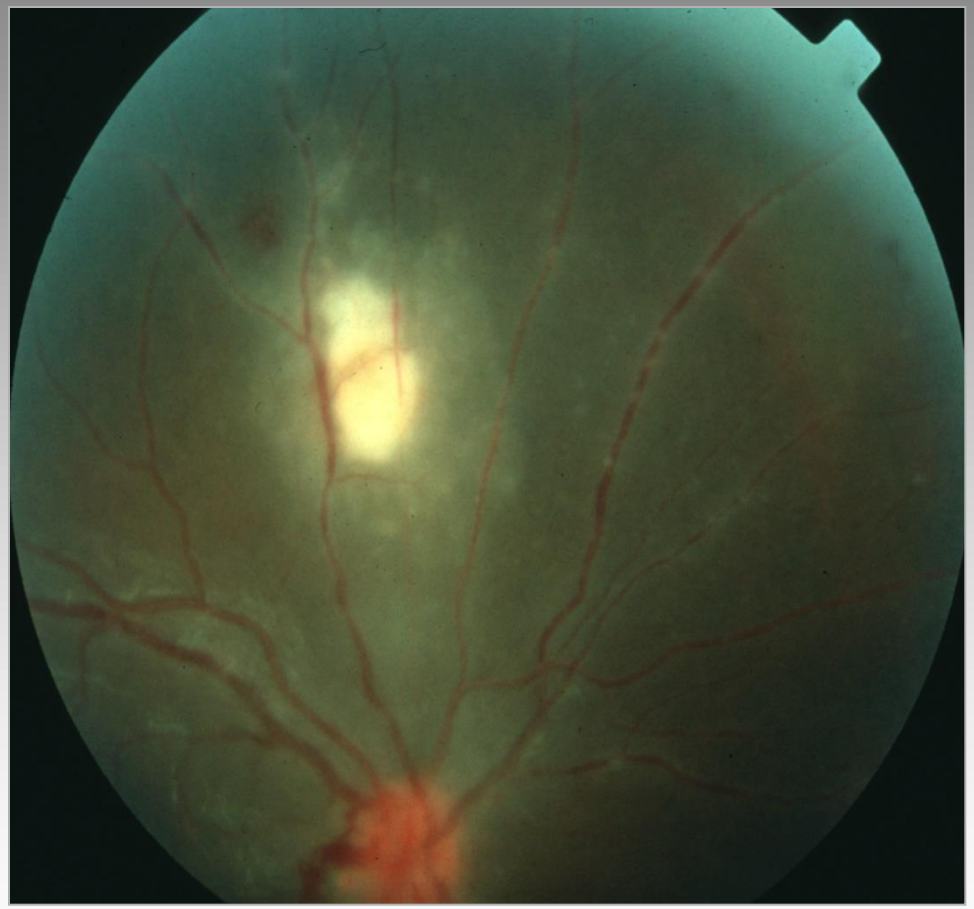


Toxoplasmic retinochoroiditis

- Most frequent post uveitis
- *Toxoplasma Gondii*
- Treatement adult:
  - Sulfadiazine 3 x 2cp à 500 mg
  - Daraprim 2 x 25mg
  - Acide folinique (leucovorin)
  - Oral prednisone

  For children cf Vaudaux B. Pediatrica

- Toxoplasmic subtypes
  - Type I, II ou III



B. Vaudaux
Pediatrica 2010

Traitement antiparasitaire

Le traitement antiparasitaire repose sur l'administration simultanée de pyriméthamine et sulfadiazine. Il doit être commencé dès connaissance du diagnostic et poursuivi jusqu'à l'âge de 12 mois.

Pyriméthamine	Posologie	Administration	Présentation
	1 mg/kg	1 jour sur 2* En 1 prise	Comprimés: 25 mg (sécables)
Sulfadiazine	**Posologie**	**Administration**	**Présentation**
	100 mg/kg (max 1 g)	1 jour sur 2* En 2 prises	Suspension non commercialisée en Suisse mais pouvant être confectionnée à partir de comprimés 500 mg pour obtenir une suspension titrant 200 mg/mL

* Pyriméthamine et sulfadiazine sont administrés le même jour

La sulfadiazine étant susceptible d'entrer en compétition avec la bilirubine pour la liaison aux protéines plasmatiques, elle est contre-indiquée chez le nouveau-né ictérique ou susceptible de le devenir. Si le traitement antiparasitaire doit être entrepris chez un nouveau-né ictérique ou de moins de 2 semaines, il est préférable de recourir à la spiramicine (en lieu et place de l'association pyriméthamine-sulfadizine) jusqu'à résolution de l'ictère ou jusqu'à la fin de la 2ème semaine.

Spiramycine	Posologie	Administration	Présentation
	50 mg/kg	Tous les jours En 2 prises	Suspension non commercialisée en Suisse mais disponible par l'intermédiaire d'une pharmacie internationale

L'action anti-acide folique de la pyriméthamine et de la sulfadiazine étant susceptible de provoquer une anémie et une neutropénie lors de traitement au long cours, il est recommandé de proposer une substitution sous forme d'acide folinique.

Acide folinique	Posologie	Administration	Présentation
	5 mg	1 jour sur 2** En 1 prise	Comprimés 15 mg

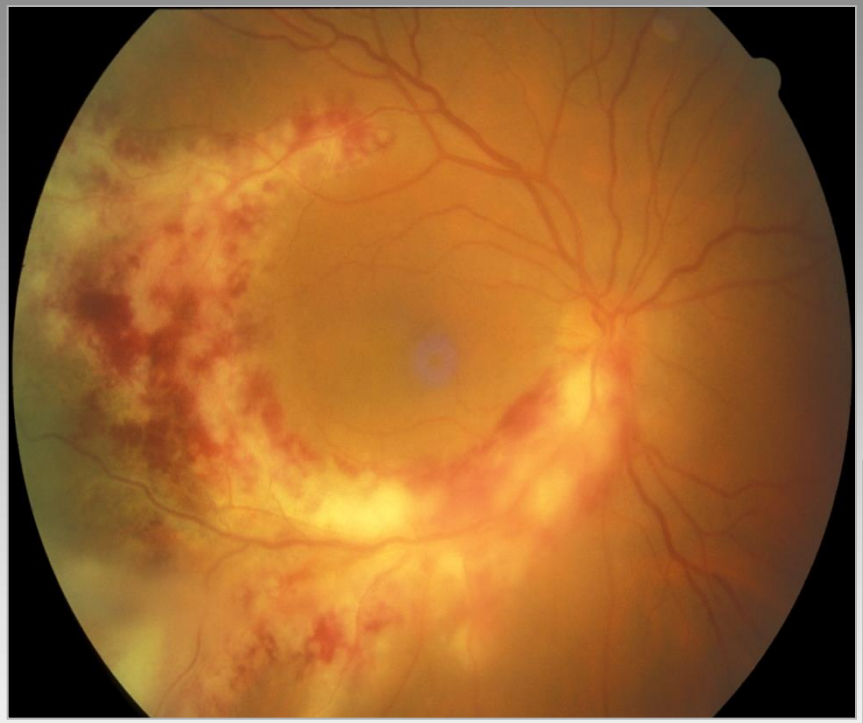
** L'acide folinique est administré en alternance avec pyriméthamine-sulfadiazine

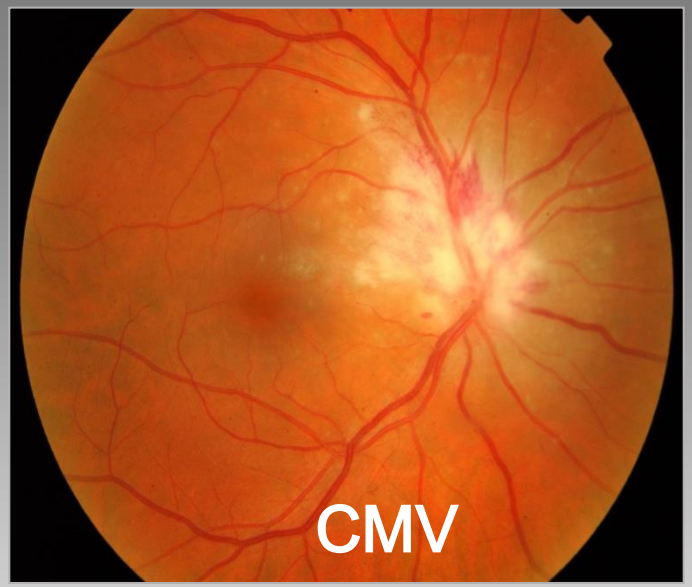
Traitement anti-inflammatoire

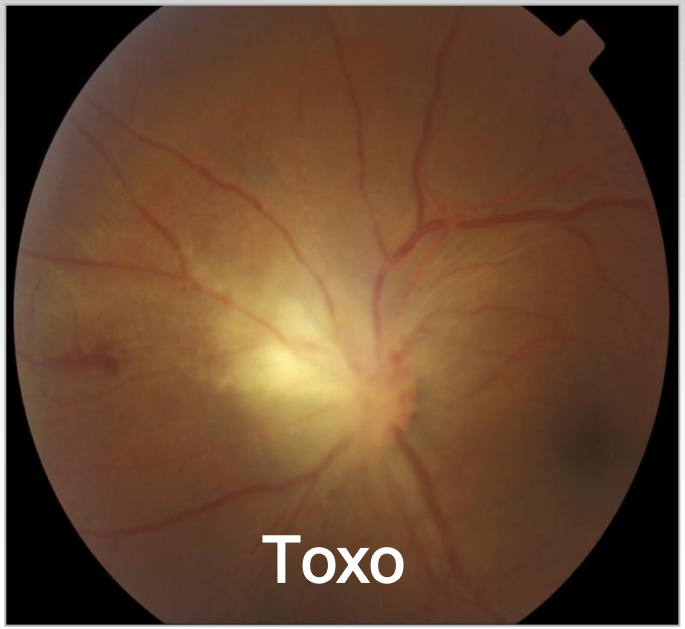
Le traitement anti-inflammatoire n'est indiqué que si l'enfant présente un ou plusieurs foyers de choriorétinite aiguë et repose sur la prednisone. Sa durée est dictée par l'état inflammatoire de la rétine et est généralement comprise entre 4 et 6 semaines.

Prednisone	Posologie	Administration	Présentation
	1 mg/kg	Tous les jours En 1 ou 2 prises	Gouttes (prednisolone) 10 mg/mL

CMV retinitis vs Toxo







Uveitis module in JIR - Cohorte

- Interdisciplinary network:

  Paediatric rheumatology – Ophthalmology

- Electronic CRF which generates automatically a pdf which is built up as a report

- Allows statistical analysis (data exportation)

- Sufficient Power to analyse orphean diseases (International cohort)

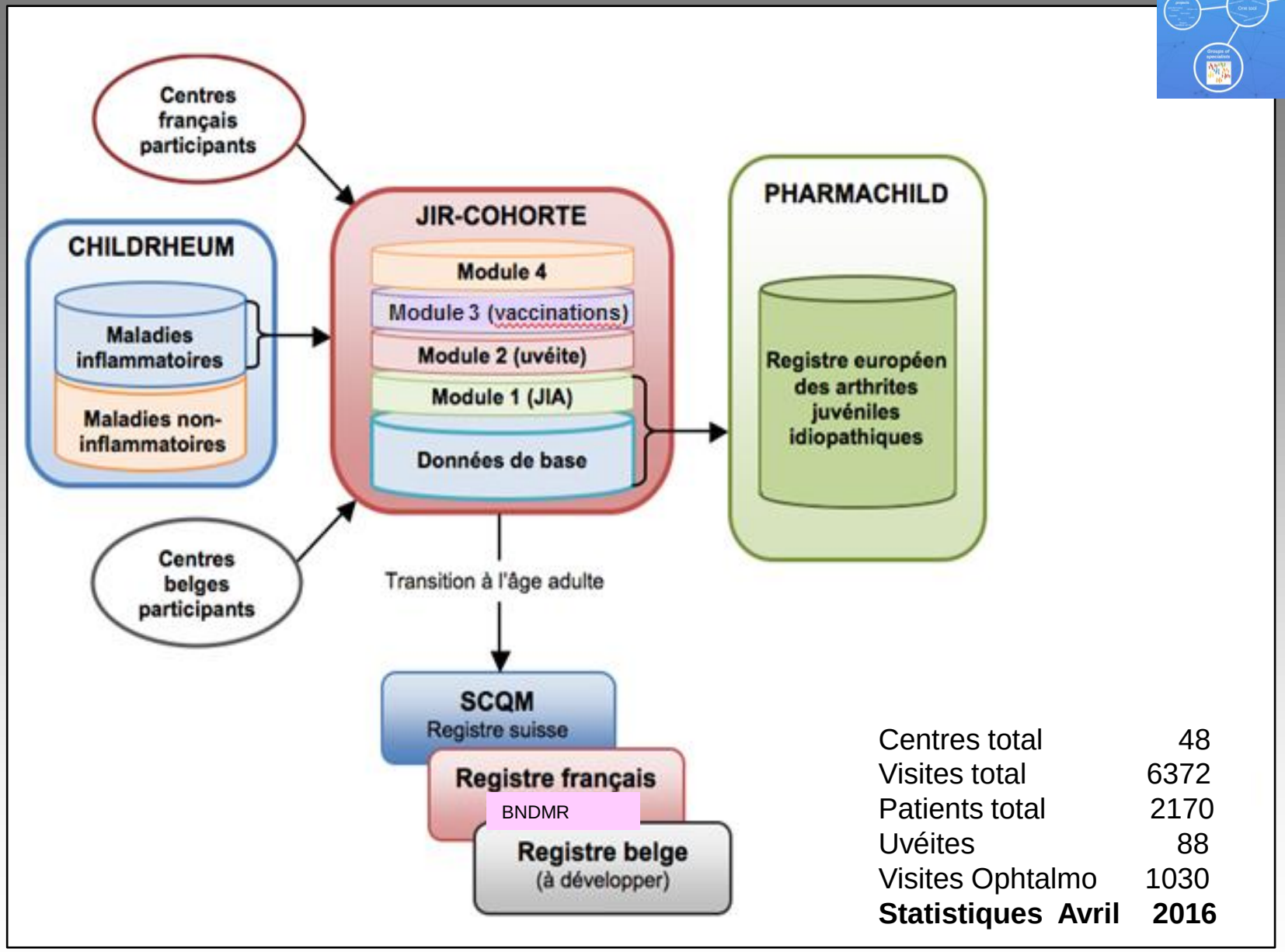
Aims

- Real time analysis of DMARDs and Biologics use
  - Relapse of ocular or systemic disease
  - Control of inflammation
  - Sequence analysis of best therapeutic
- Averse events notification and statistics
- Study of the mechanisms of uveitis : Uveitis biobank since 2000 with (collection of more than 1500 serum and DNA)

JIR-Cohorte and uveitis

- Orphean disease
- Online database Https://jircohorte.seantis.ch/login
  - Similar platform SCQM / rhumatologie adulte
    - Database for biological therapies in Switzerland
    - Secure Server (medical data)
    - Development by SEANTIS (Lucern)

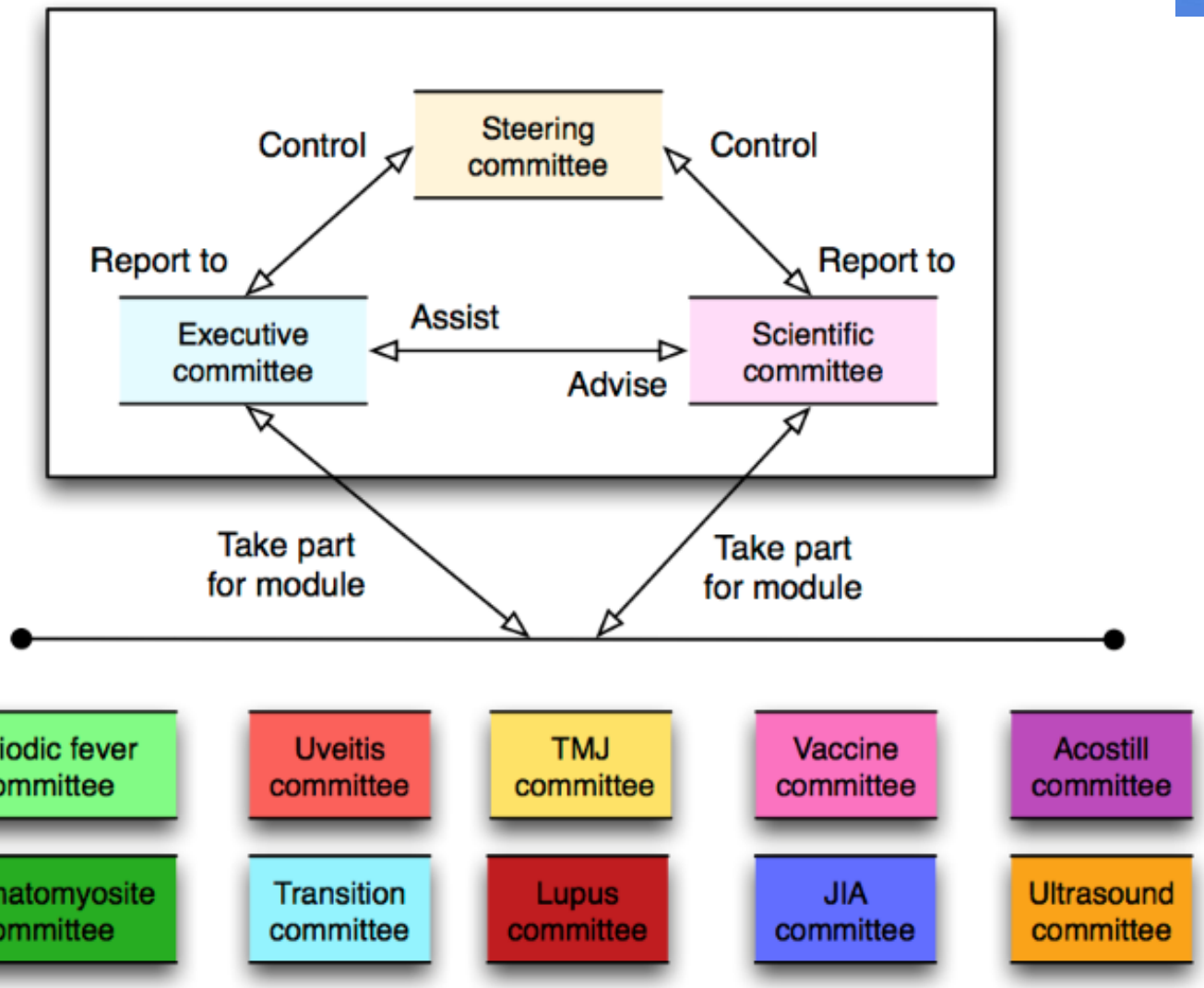
  - Basis : Pediatric rheumatologists
    - Suisse (Bâle, Zurich, Berne, Lausanne, Genève) + Lyon, Paris, Belgique



Centres total	48
Visites total	6372
Patients total	2170
Uvéites	88
Visites Ophtalmo	1030
Statistiques Avril	**2016**

JIR-cohorte

JIRcohorte committees



Online data collection (CRF)

Tonometry right eye

9 mmHg

Tonometry left eye

10 mmHg

Cornea right eye

- [x] Claire
- [ ] Précipités endothéliaux fins
- [ ] Précipités endothéliaux gran
- [ ] Kératopathie en bandelettes
- [ ]

Cornea left eye

- [x] Claire
- [ ] Précipités endothéliaux fins
- [ ] Précipités endothéliaux gran
- [ ] Kératopathie en bandelettes
- [ ]

Grading anterior chamber cells right eye

☐ 0 ☑ 0.5+ ☐ 1+ ☐ 2+ ☐ 3+ ☐ 4+

Grading anterior chamber cells left eye

☐ 0 ☑ 0.5+ ☐ 1+ ☐ 2+ ☐ 3+ ☐ 4+

Grading anterior chamber flare right eye

☑ 0 ☐ 1+ ☐ 2+ ☐ 3+ ☐ 4+

Grading anterior chamber flare left eye

☐ 0 ☑ 1+ ☐ 2+ ☐ 3+ ☐ 4+

Grading vitreous cells right eye

☑ 0 ☐ 0.5 ☐ 1 ☐ 2 ☐ 3 ☐ 4

Grading vitreous cells left eye

☑ 0 ☐ 0.5 ☐ 1 ☐ 2 ☐ 3 ☐ 4

Grading vitreous opacities right eye (NEI vitreous haze grading scale)

☑ 0 ☐ trace ☐ 1+ ☐ 2+ ☐ 3+ ☐ 4+

Grading vitreous opacities left eye (NEI vitreous haze grading scale)

☑ 0 ☐ trace ☐ 1+ ☐ 2+ ☐ 3+ ☐ 4+

Snowbanking right eye

☑ Non ☐ Oui

Snowbanking left eye

☑ Non ☐ Oui

Snowballs right eye

Snowballs left eye

Therapies
Shared between Ped rhumatol / uveitis

1. Profil ⓘ | 2. Historique | 3. Consultation | 4. Investigations | 5. Evaluation | **6. Plan** ⓘ | 7. Rapport

Médicaments ⓘ | Effets secondaires | Recommandation

+ Ajouter traitement

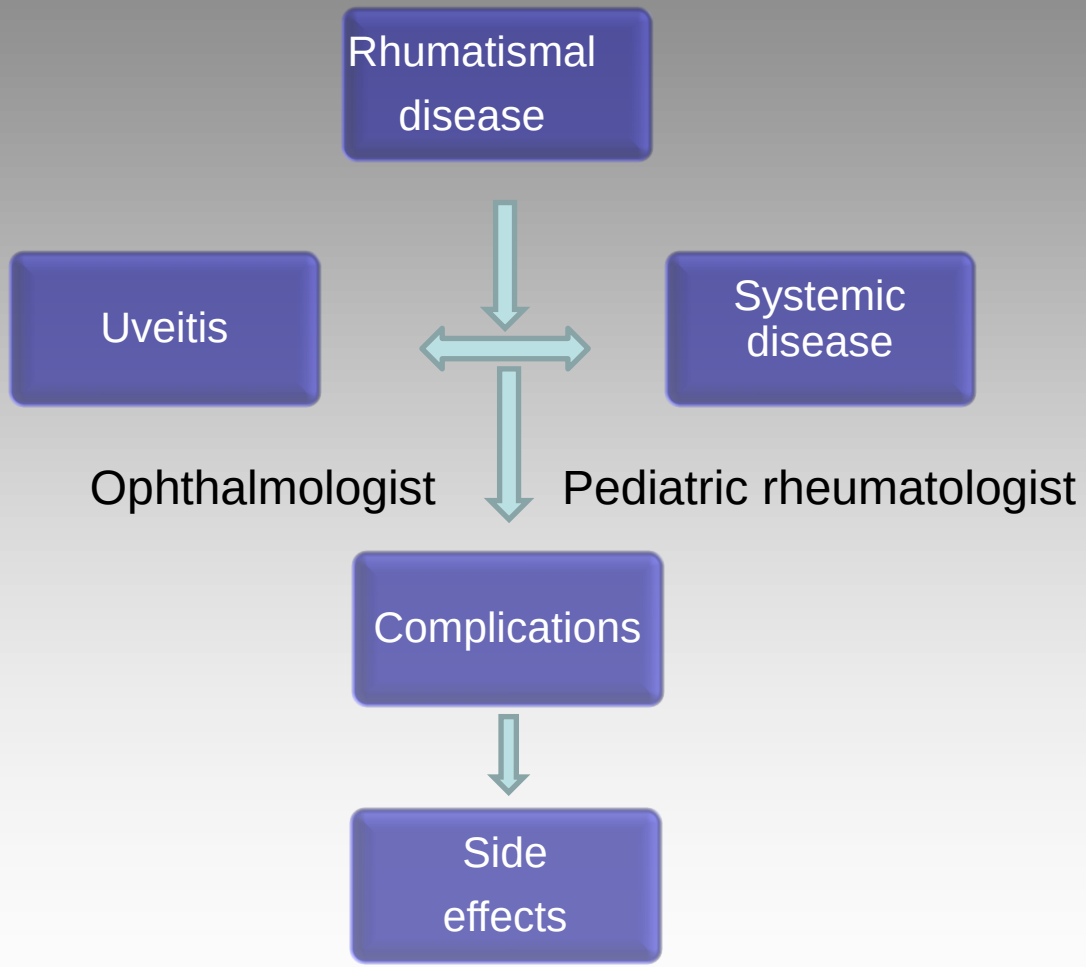
Traitement actuel

Médicament	Début	Ajustement	Dose	Fréquence	Route	
Humira	5.1.2015		40 mg	Toutes les 2 semaines	SC	Ajuster ✕ Arrêter
Methotrexate	1.8.2013	20.1.2016	10 mg	Chaque semaine	PO	Ajuster ✕ Arrêter
acide folique	16.9.2015		5 mg	Chaque semaine	PO	Ajuster ✕ Arrêter

Traitement interrompu

Médicament	Début	Ajustement	Dose	Fréquence	Route	Fin	Raison de l'arrêt
timoptic XE 0.5 ⓘ	16.9.2015		1 Goutte	Chaque jour	OU	9.12.2015	

Conclusions Module uveitis module => real time analysis of ocular and systemic complications



Milestones of the project

2010
- Uveitis data collections validated by the Swiss group of paediatric rheumatology / Swiss specialists in uveitis

2015
- Online
  Bâle, Berne, Zurich, Genève

2016
- Extension Europe (Lyon, Paris, Bruxelles)
- 2170 patients ; 2500 données
  88 pediatric uveitis included / 150 with transition module in Lausanne

2017
- Futur : Extension European project ⇨ Adult module