

AAPOS Pediatric Uveitis Committee:

How (Not) to Manage Uveitis: Lessons Learned from Complicated Cases

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The goal of this workshop is to provide updates in the medical and surgical management of children with uveitis using “real life” cases. The practice of medicine is humbling, but willingness to learn, reflect and share these lessons will improve uveitis care.

Section 1: Diagnosis

10% of Uveitis is secondary to infectious causes, but proper infectious diagnosis requires recognition of risk factors and proper testing. The following is a guide to differential infectious from non-infectious uveitis. Uveitis from Non-Infectious Uveitis

A. Infectious Causes based on Anatomic Location

ANTERIOR	INTERMEDIATE	POSTERIOR/PANUVEITIS
<i>ALWAYS RULE OUT SYPHILLIS AND TB</i>	<i>ALWAYS RULE OUT SYPHILLIS AND TB</i>	<i>ALWAYS RULE OUT SYPHILLIS AND TB</i>
Herpes (HSV/VZV/CMV)	Lyme	Toxoplasmosis
Lyme	Toxocariasis	Toxocariasis
	Bartonella	Bartonella
		Herpes (HSV/VZV/CMV)
		Rubella, Rubeola
		TORCH, plus Zika and LCMV

B. History that is suspicious for infectious cause

1. Pets (dogs/puppies, Toxocariasis; Cats, toxoplasmosis and Bartonella)
2. Travel and geographic region (Lyme, Histoplasmosis)
3. Immune suppressed status (HIV, Transplant patients)

C. Anterior segment exam findings suggestive of viral etiology

1. Recurrent or chronic unilateral, non-alternating disease
2. High IOP upon presentation
3. Iris Transillumination defects
4. Stellate KP (small central/paracentral KP)
5. Keratitis/Endothelitis

Suggested Resource:

Jap A, Chee, S; Viral Anterior Uveitis: Diagnosis and Management; AAO Focal Points: Clinical Practice Perspectives 2016.

Stepladder Approach To Uveitis Treatment

3

**Biologic Response Modifiers
(Biologics)**

2

**Conventional IMT:
Methotrexate (MTX) –preferred
Other: Cyclosporine, Leflunomide ,
Mycophenolate**

1

**Topical corticosteroids
+/- Periocular steroids
+/- Systemic steroids (< 3 months)**

For severe sight-threatening disease, Simultaneous initiation of MTX (or other DMARD) and conventional TNFalpha inhibitor may be considered

Errors in medication administration due to patient confusion are large source of medication failure. Taking the time to create clear and detailed patient instructions will improve patient compliance and outcomes.

Attached is an example of a patient instruction table.

If you copy and paste this into a smartphrase creator in EPIC, you can pull it up on the AVS through a dot phrase.

Prednisolone acetate – pink or white cap



1 drop *** times per day in *** eye

Durezol (difluprednate) – pink cap



1 drop *** times per day in *** eye

Cyclopentolate (Cyclogyl) – red cap



1 drop *** times per day in *** eye

Cosopt (dorzolamide-timolol) – navy cap

1 drop *** times per day in *** eye



Latanoprost – teal cap

1 drop *** times per day in *** eye



Timolol – yellow cap

1 drop *** times per day in *** eye



Section 3: Surgical management

Complications of cataract surgery in patient with uveitis include:
Cyclitic membranes, Hypotony, Phthisis, Glaucoma, Macular edema

Best Practice to avoid these complications include

1. Wait for surgery until uveitis is controlled. Ideally uveitis controlled for at least 3 months, but if patient can wait longer, wait longer.
2. Relative contraindication for IOL
 - a. Patients less than 4 years old
 - b. Pre-surgical hypotony
 - c. IOL complications in fellow eye
 - d. Shallow AC
3. Make a clear plan to control peri-operative inflammation
 - a. Consider oral steroids and topical steroids starting pre-op
 - b. Coordinate with rheumatology and consider timing of surgery related to patient's immune suppressive dosing schedule

Key Treatment References & Resources:

- American College of Rheumatology (ACR)/Arthritis Foundation Guideline for the Screening, Monitoring, and Treatment of Juvenile Idiopathic Arthritis-Associated Uveitis.

- Applies to JIA-uveitis (or by extension idiopathic JIA-like, chronic anterior uveitis)

- Reference: **Angeles-Han ST**, Ringold S, Beukelman T, Lovell D, Cuello CA, Becker ML, Colbert RA, Feldman BM, Holland GN, Ferguson PJ, Gewanter H, Guzman J, Horonjeff J, Nigrovic PA, Ombrello MJ, Passo MH, Stoll ML, Rabinovich CE, Sen HN, Schneider R, Halyabar O, Hays K, Shah AA, Sullivan N, Szymanski AM, Turgunbaev M, Turner A, Reston J. 2019 American College of Rheumatology/Arthritis Foundation Guideline for the Screening, Monitoring, and Treatment of Juvenile Idiopathic Arthritis-Associated Uveitis. *Arthritis Care Res (Hoboken)*. 2019 Jun;71(6):703-716. doi: 10.1002/acr.23871. Epub 2019 Apr 25. PMID: 31021540; PMCID: PMC6777949. Full text link: <https://onlinelibrary.wiley.com/doi/full/10.1002/acr.23871>

- Consensus-based recommendations for the management of uveitis associated with juvenile idiopathic arthritis: The SHARE (The Single Hub and Access point for pediatric Rheumatology in Europe) Initiative.

- -Overlapping features with ACR guidelines above, some differences

- Reference: Constantin et al., *Ann Rheum Dis* 2018; 77 (8); 1107-1117.

- Full text link: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6059050/>

- Childhood Arthritis and Rheumatology Research Alliance (CARRA) Consensus Treatment Plans (CTPs) for Juvenile Idiopathic Arthritis-Associated and Idiopathic Chronic Anterior Uveitis

- Angeles-Han ST**, Lo MS, Henderson LA, **Lerman MA**, Abramson L, **Cooper AM**, Parsa MF, Zemel LS, Ronis T, Beukelman T, Cox E, Sen HN, Holland GN, Brunner HI, Lasky A, Rabinovich CE; Juvenile Idiopathic Arthritis Disease-Specific and Uveitis Subcommittee of the Childhood Arthritis Rheumatology and Research Alliance. Childhood Arthritis and Rheumatology Research Alliance Consensus Treatment Plans for Juvenile Idiopathic Arthritis-Associated and Idiopathic Chronic Anterior Uveitis. *Arthritis Care Res (Hoboken)*. 2019 Apr;71(4):482-491. doi: 10.1002/acr.23610. PMID: 29806733; PMCID: PMC6261704.- Applies to the design of prospective research to compare effectiveness of treatment

- These are not treatment guidelines

- Full text link: <https://www.rheumatology.org/Portals/0/Files/JIA-Uveitis-Guideline-2019.pdf>

- Tirelli F, Shafer BM, **Davidson SL**, **Lerman MA**. Immunomodulation and TNF- α inhibition for tubulointerstitial nephritis and uveitis syndrome: a case series. *J AAPOS*. 2021 Oct;25(5):267.e1-267.e6. doi: 10.1016/j.jaapos.2021.03.017. Epub 2021 Sep 29. PMID: 34600106.

- Margaret Reynolds, MD; Wendy Smith, MD; Knights of Templar: Pediatric Ophthalmology Education Center: Pediatric

- Intermediate Uveitis <https://www.aao.org/disease-review/pediatric-intermediate-uveitis>

- Wang, RC Intermediate Uveitis/Pars Planitis, *AAO Focal Points* Feb 2017.

