



Oculoplastics Disorders

Primary Acquired
Nasolacrimal Duct
Obstruction

Orbital Inflammatory
Syndrome

INSIDE:

- Two Common Oculoplastics Disorders
- Diagnosis and Treatment
- Referral Guidelines



MASSACHUSETTS
EYE AND EAR



MASSACHUSETTS
GENERAL HOSPITAL

eye Insights[™]

Dear Colleagues,

As ophthalmologists, one of the most exciting facets of our profession is our role as problem-solvers. Some of our patients have typical complaints and clear signs pointing to their pathologies, while others have complex, multifactorial problems, often connected to underlying systemic conditions that require all of our clinical skills and knowledge to accurately diagnose and treat.

In this issue of *eye Insights*, we take a close look at two common oculoplastics disorders seen in ophthalmology practice: primary acquired nasolacrimal duct obstruction (PANDO) and orbital inflammatory syndrome (OIS). Inside, you'll find key information about how PANDO and OIS are diagnosed and treated, guidance on when to refer a patient, and further reading.

Keep in mind that ophthalmic plastic surgery is a unique specialty that relies on a surgeon's technical ability and artistic skill. For a list of doctors that specialize in this type of surgery, please visit the website for the American Society of Ophthalmic Plastic & Reconstructive Surgery.

We hope you find this issue of *eye Insights* useful in your practice. Back issues are available online at masseyeandear.org. If you have questions or comments, please email us at eyeinsights@meei.harvard.edu.



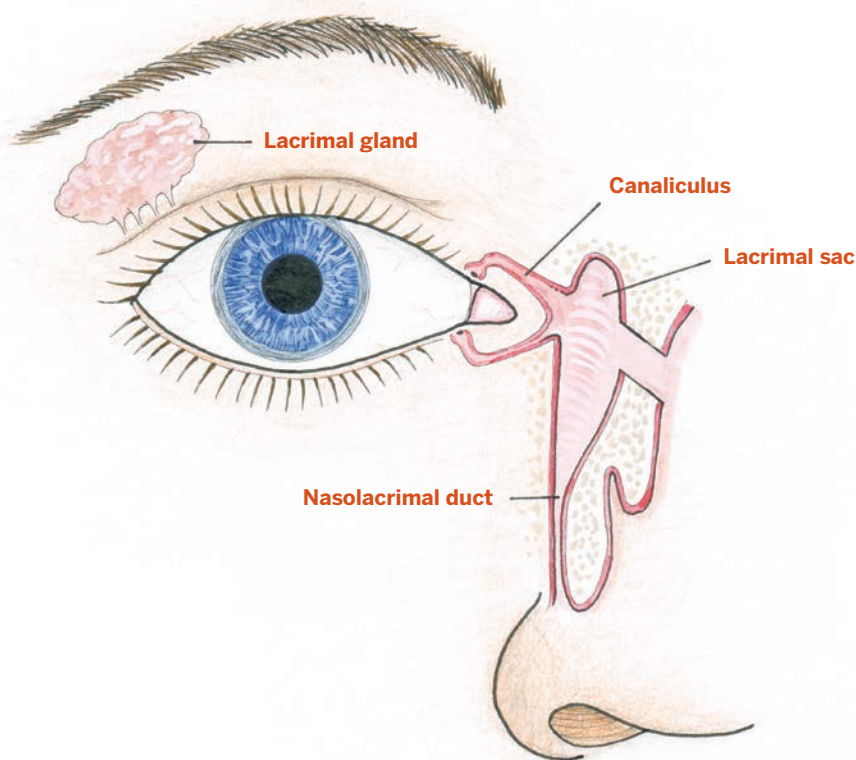
A handwritten signature in blue ink that reads "Joan W. Miller". The signature is fluid and cursive, with the first letters of the first and last names being capitalized and prominent.

Joan W. Miller, MD

*David Glendenning Cogan
Professor of Ophthalmology and
Chair, Department of Ophthalmology
Harvard Medical School*

*Chief of Ophthalmology,
Massachusetts Eye and Ear and
Massachusetts General Hospital*

*Ophthalmologist-in-Chief,
Brigham and Women's Hospital*



TWO COMMON OCULOPLASTICS DISORDERS

Primary Acquired Nasolacrimal Duct Obstruction (**PANDO**)

- PANDO is a condition in which chronic inflammation and scarring block the tear drainage system without another identifiable cause for the obstruction, such as trauma, tumor, etc. Patients with PANDO usually seek help when tears overflow. Patients with obstructions are also at risk for infection of the nasolacrimal system.

Orbital Inflammatory Syndrome (**OIS**)

- OIS refers to noninfectious inflammation of the orbital tissues—the lacrimal gland (dacryoadenitis), extraocular muscles (myositis), and sclera (scleritis)—or diffuse inflammation and/or a combination of any of these tissues. The inflammation may be idiopathic or may be secondary to a bona fide inflammatory condition, such as sarcoidosis, granulomatosis with polyangiitis, IgG4-related disease, and others.
- Patients with OIS often seek help because their eyelids and the area around their eyes are red, swollen, and painful. The eye may be protruding. Vision problems, including double vision and a decrease of vision, can occur.



Daniel R. Lefebvre, MD, FACS

Assistant Professor of Ophthalmology,
Harvard Medical School

Ophthalmic Plastic Surgery Service
at Mass. Eye and Ear

PANDO

ASK THE EXPERT



Suzanne Freitag, MD

Associate Professor of Ophthalmology,
Harvard Medical School

Director, Ophthalmic Plastic Surgery
Service, and Co-Director, Center for
Thyroid Eye Disease and Orbital Surgery
at Mass. Eye and Ear

OIS

- The gold standard for diagnosis is irrigation of saline through the tear drainage system via a lacrimal cannula through the punctum and canaliculi. Passage of saline through the system into the nasal cavity confirms that the system is patent. Reflux from the opposing punctum indicates an obstruction.
- Diagnosis of acute or chronic infection of the tear drainage system confirms a diagnosis of PANDO. This is diagnosed by visualizing an enlarged and inflamed lacrimal sac or discharge coming from the puncta when pressure is applied on the lacrimal sac.
- Maxillofacial CT scan is considered for certain cases—such as young adults, those with a history of maxillofacial trauma or malignancy, and those with severe chronic rhinosinusitis—to determine if concurrent functional sinus surgery may be appropriate.
- Nasal endoscopy may reveal problems in the nasal cavity requiring surgery, such as a deviated nasal septum, chronic rhinosinusitis, concha bullosa (air pocket in the middle concha), tumor, or scarring.
- Another test for PANDO is to trace the movement of dye through the lacrimal drainage system in one of two ways: 1) dacryocystography, where dye is injected in the tear duct and traced with X-ray and/or CT scan, or 2) dacryoscintigraphy, where a radiotracer eye drop is placed in the conjunctival cul-de-sac and its movement is recorded over time through a series of gamma camera images.

DIAGNOSIS

- As part of a thorough history, it's important to ask about the use of medications associated with OIS, such as bisphosphonates taken for the treatment of osteoporosis.
- A comprehensive ophthalmic evaluation is performed. Pain can be an important symptom of OIS, particularly in myositis and dacryoadenitis. It's important to rule out infectious orbital cellulitis, which in the vast majority of cases originates from paranasal sinusitis.
- Most often an orbital CT scan with contrast will clearly image the orbit and paranasal sinuses and aid in the diagnosis. A CT scan is typically preferred as the initial imaging modality of choice as it is quickly obtained, has high spatial resolution, and shows bony anatomy in detail that is important in cases of infection or malignancy. MRI can subsequently be obtained if needed to augment the workup.
- An orbital biopsy helps to confirm the diagnosis of OIS in cases in which other

etiologies are not found with laboratory testing or other less invasive testing. The histopathology may demonstrate specific signs, such as granulomatous inflammation (as in either sarcoidosis or granulomatosis with polyangiitis) or a high number of IgG4 plasma cells (as in IgG4-related disease).

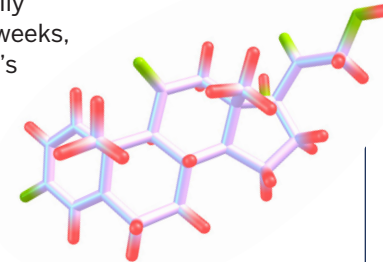
To rule out infection or systemic disease, clinicians might order these additional tests:

- A complete blood count and basic metabolic panel
- Thyroid studies: free T4, T3, thyroid peroxidase antibody (TPO), thyroid-stimulating hormone (TSH), thyroid-stimulating immunoglobulin (TSI), and thyrotrophin-binding inhibitor immunoglobulin (TBII)
- Tests for antinuclear antibodies, antineutrophil cytoplasmic antibodies, angiotensin converting enzyme, rapid plasma reagent, rheumatoid factor, and quantiferon gold (T-spot) to rule out tuberculosis.

- Dacryocystorhinostomy (DCR) surgery is the primary treatment for PANDO. Surgeons direct the lacrimal sac to drain into the nasal cavity in a slightly different location, bypassing the obstruction. DCR resolves tearing and infection in more than 90% of patients.
- DCR surgery may be performed traditionally via a small skin incision by the inner lower eyelid area that heals very well, or via an intranasal endoscopic approach. These techniques yield equivalent success rates, and either technique may be appropriate based on surgeon experience and potential need for other intranasal procedures (such as septoplasty or sinus surgery).

TREATMENT OPTIONS

- In very mild cases, observation may often be the first choice.
- Oral NSAIDs are commonly used as first-line therapy in mild cases.
- Systemic corticosteroid therapy (oral prednisone) is used in many cases of typical dacryoadenitis or myositis. A taper is typically performed over six to eight weeks, and is tailored to the patient's improvement in signs and symptoms.
- Second-line immunosuppressive therapies (cyclosporine, cyclophosphamide, tacrolimus, methotrexate, rituximab, and others) are often used for OIS linked to systemic conditions as well as for sclerosing OIS.
- Other options include orbital radiation therapy and, very rarely, surgical debulking.



- All patients diagnosed with PANDO should be referred to an oculoplastic surgeon with expertise in lacrimal surgery. Ophthalmologists are also encouraged to refer PANDO suspects who require advanced testing for an accurate diagnosis.

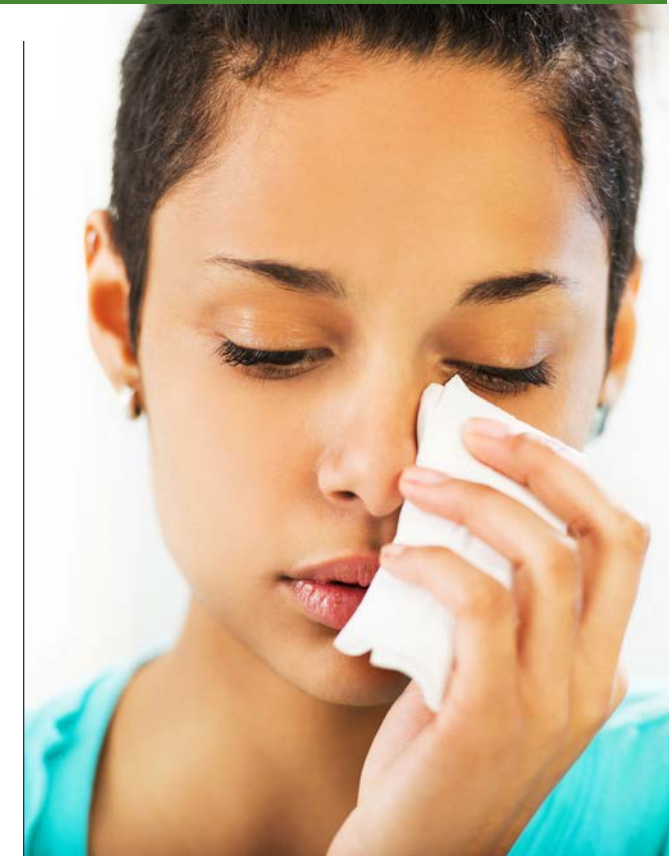
REFERRAL GUIDELINES

- Any patient with orbital inflammation should be referred to an orbit specialist. Treatment can be complex, encompassing therapies for systemic disease, as well as corticosteroids, surgery, and radiation. The orbit specialist typically coordinates care with other specialties, such as rheumatology, to meet all of the patient's needs.

- Woog JJ. The incidence of symptomatic acquired lacrimal outflow obstruction among residents of Olmsted County, Minnesota, 1976–2000 (An American Ophthalmological Society Thesis). *Trans Am Ophthalmol Soc.* 2007;105: 649-666.

FURTHER READING

- Lefebvre DR, Reinshagen KL, Yoon MK, et al. An 18-year-old man with diplopia and proptosis of the left eye. *N Engl J Med.* 2018;379(25):2452-2461.
- Lefebvre DR, Mandeville JT, Yonekawa Y, et al. A case series and review of bisphosphonate-associated orbital inflammation. *Ocul Immunol Inflamm.* 2016;24(2):134-139.
- Kubota T. Corticosteroids or biopsy for idiopathic orbital inflammation. *Surv Ophthalmol.* 2017;62(2):253-255.



eye Insights™ Spring 2019

Editor-in-Chief:
Joan W. Miller, MD

Managing Editor:
Matthew F. Gardiner, MD

Clinical Advisory Group:
Deeba Husain, MD
Ankoor S. Shah, MD, PhD

Contributors:
Suzanne Freitag, MD
Daniel R. Lefebvre, MD, FACS

Design:
J Sherman Studio

Ophthalmology Communications:
Suzanne Ward, director
Rosa Rojas, manager
Elizabeth DiVito, editor

Illustration:
Eye diagram by
Laurel Cook Lhowe

Photography:
Licensed from istock.com
Tulips by jimfeng
Background by IrisImages
Prednisone molecule by ollaweila
Woman with tears by skynesher

Published biannually, *eye Insights* offers the ophthalmology community best practice information from Mass. Eye and Ear specialists.

We welcome your feedback. Send comments to:
eyeinsights@meei.harvard.edu

