

## Supplementary Online Content

Mombaerts I, Bilyk JR, Rose GE, et al; for the Expert Panel of the Orbital Society. Consensus on diagnostic criteria of idiopathic orbital inflammation using a modified Delphi approach. *JAMA Ophthalmol*. Published online June 1, 2017. doi:10.1001/jamaophthalmol.2017.1581

**eTable 1.** Questionnaire and Scores of the First Delphi Round

**eTable 2.** Questionnaire and Scores of the Second Delphi Round

**eFigure.** The Initial Diagnostic Steps (With or Without Serology) Among Experts (N = 27), for Suspected IOI in the Lacrimal Gland, Muscle, or Orbital Fat

This supplementary material has been provided by the authors to give readers additional information about their work.

**eTable 1.** Questionnaire and Scores of the First Delphi Round

	<b>Median</b>	<b>IQR</b>	<b>No. Positive Responders*</b>	<b>Consensus on</b>
<b>The following clinical and radiologic imaging findings are indicative of idiopathic dacryoadenitis:</b>				
Age between 10 and 75 years	4	1	18 (67%)	Agreement
No history of lacrimal-gland related systemic disease	5	1	24 (89%)	Agreement
Acute or subacute onset	4	0	24 (89%)	Agreement
Duration > 6 weeks	3	2	11 (41%)	None
Orbital pain	4	1	24 (85%)	Agreement
Superotemporal (epi)scleritis	3	2	10 (37%)	None
Unilateral	4	1	17 (63%)	Agreement
Diffuse lacrimal gland swelling	4	1	23 (85%)	Agreement
Irregular margins of the lacrimal gland mass	3	1	12 (44%)	Neutral
Contrast enhancement	4	1	24 (89%)	Agreement
Intact lacrimal bone	5	1	25 (93%)	Agreement
Extension in adjacent extraocular muscle, eyelid and/or extraconal fat	3	1	13 (48%)	Neutral
Normal ipsilateral paranasal sinus	4	2	20 (74%)	None
Absence of infra- and supraorbital nerve enlargement	4	1	21 (78%)	Agreement
<b>The following clinical and radiologic imaging findings are indicative of idiopathic orbital myositis:</b>				
Age between 10 and 60 years	4	1	18 (67%)	Agreement
Acute or subacute onset	4	1	24 (89%)	Agreement
No history of thyroid disease	4	4	24 (89%)	Agreement
Orbital pain	5	1	25 (93%)	Agreement
Pain on eye movements	5	1	26 (96%)	Agreement
Limited eye movements	4	1	23 (85%)	Agreement
(Epi)scleritis at insertion of involved muscle	4	2	19 (70%)	None
Any extraocular muscle affected	4	1	25 (93%)	Agreement
Diffuse extraocular muscle swelling	4	0	24 (89%)	Agreement

Anterior tendon involved	4	0.5	20 (74%)	Agreement
Contrast enhancement	4	1	23 (85%)	Agreement
Normal ipsilateral paranasal sinus	4	1.75	17 (63%)	None
Absence of supra- and infraorbital nerve enlargement	4	1	22 (81%)	Agreement
Absence of superior ophthalmic vein enlargement	4	1.5	19 (70%)	None
<b>The following clinical and radiologic imaging findings are indicative of idiopathic orbital fat inflammation :</b>				
Age between 10 and 75 years	4	1	14 (67%)	Agreement
No history of orbital-related systemic disease	4	1	23 (92%)	Agreement
Acute or subacute onset	4	1	23 (92%)	Agreement
Orbital pain	4	1	20 (80%)	Agreement
Unilateral	4	1	16 (64%)	Agreement
Diffuse orbital mass	4	1	17 (71%)	Agreement
Irregular margins of the orbital fat mass	4	1	19 (79%)	Agreement
Contrast enhancement	4	0.5	22 (92%)	Agreement
Adjacent extraocular muscle or lacrimal gland invasion	4	1	16 (67%)	Agreement
Absence of supra- and infraorbital nerve enlargement	4	2	16 (63%)	None
Normal ipsilateral paranasal sinus	4	1	16 (64%)	Agreement
Intact orbital bones	4	1	23 (93%)	Agreement
<b>The following histopathological and immunohistochemical findings are indicative of idiopathic dacryoadenitis :</b>	<b>Median</b>	<b>IQR</b>	<b>No. Positive Responders*</b>	<b>Consensus on</b>
Fibrosis	4	1	13 (65%)	Agreement
Lymphoplasmacytic infiltrate	4	1	24 (96%)	Agreement
Lymphoid follicles with reactive germinal centers	4	2	13 (52%)	None
Acini and/or duct destruction	3	1.5	9 (38%)	None
Absence of granulomatous inflammation, necrosis and/or vasculitis	5	1	19 (79%)	Agreement
Absence of epimyoeptithelial islands	4	1.25	18 (75%)	Agreement
If plasma cells stain IgG4	3	1	10 (38%)	Neutral

positive, the IgG4+/IgG ratio is $\leq 40\%$ , or the count is $\leq 100/\text{hpf}$				
<b>The following histopathological and immunohistochemical findings are indicative of idiopathic orbital myositis :</b>				
Absence of fibrosis	3	2	9 (41%)	None
Lymphoplasmacytic infiltrate	4	1	25 (100%)	Agreement
Lymphoid follicles with reactive germinal centers	3	2	8 (32%)	None
Degeneration of myofibers	3	1	9 (38%)	Neutral
Absence of granulomatous inflammation, necrosis and/or vasculitis	4	1.5	17 (68%)	None
If plasma cells stain IgG4-positive, the IgG4+/IgG ratio is $\leq 40\%$ , or the count is $\leq 10/\text{hpf}$	4	1	10 (40%)	None
<b>The following histopathological and immunohistochemical findings are indicative of idiopathic orbital fat inflammation :</b>				
Fibrosis	4	1	12 (57%)	None
Lymphoplasmacytic infiltrate	4	1	23 (96%)	Agreement
Lymphoid follicles with reactive germinal centers	4	1.25	13 (54%)	None
Absence of granulomatous inflammation, necrosis and/or vasculitis	4	1.25	18 (75%)	None
If plasma cells stain IgG4-positive, the IgG4+/IgG ratio is $\leq 40\%$ , or the count is $\leq 10/\text{hpf}$	4	1	11 (46%)	None

\*a positive responder implies a 4 or 5 Likert-rating  
IQR = interquartile range; IgG4 = immunoglobulin G4.

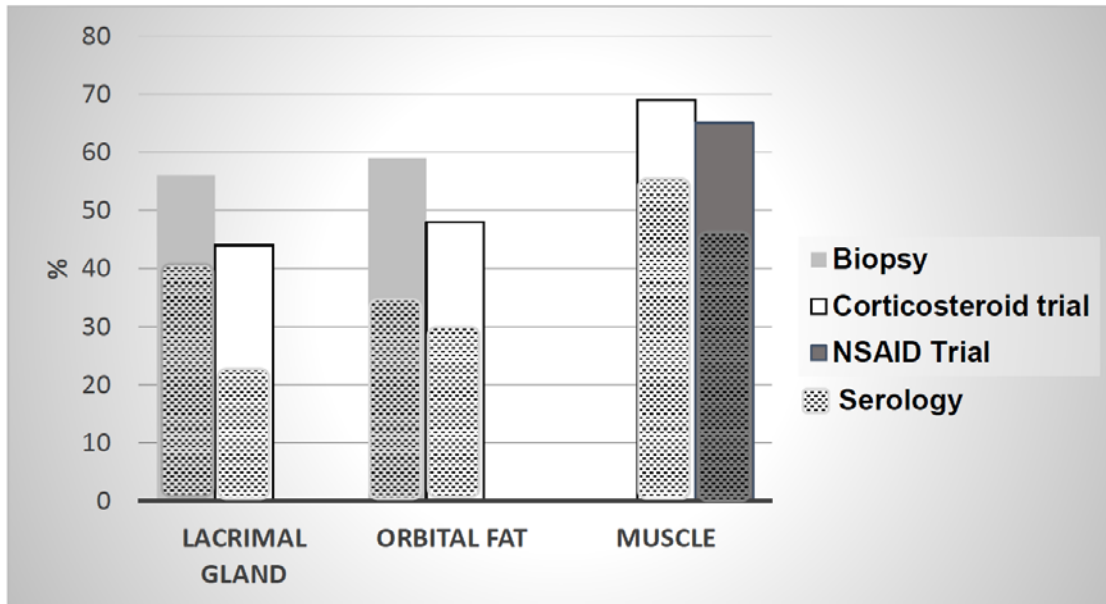
**eTable 2. Questionnaire and Scores of the Second Delphi Round**

	Median	IQR	No. Positive Responders*	Consensus on
<b>The most accurate assessment of non-myositic IOI is based on clinical indicators, MRI or CT studies, selected normal laboratory findings, and incisional biopsy.</b>	5	0	24 (89%)	Agreement
<b>The most accurate assessment of non-myositic IOI is based on clinical indicators, MRI or CT studies, selected normal laboratory findings, and corticosteroid responsiveness.</b>	3	2	12 (44%)	None
<b>Diagnostic confirmation by biopsy in non-myositic IOI is always recommended, for the exclusion of malignancy and other IOI mimics.</b>	4	2	17 (63%)	None
<b>The most accurate assessment of myositic IOI is based on clinical indicators, MRI or CT studies, selected normal laboratory findings, and corticosteroid responsiveness.</b>	4	1	22 (85%)	Agreement
<b>Diagnostic confirmation by biopsy in myositic IOI is always recommended, for the exclusion of malignancy and other myositis mimics.</b>	4	3	15 (56%)	None
<b>Re-rating of round I and newly suggested indicators of IOI</b>				
Prodromal ache shortly before eyelid swelling	3	0.5	7 (26%)	Neutral
S-shaped upper eyelid ptosis (in idiopathic dacryoadenitis)	3	1	13 (48%)	Neutral
Red demarcation line under the brow (in nonmyositic IOI)	3	0.5	5 (19%)	Neutral
No process outside the orbit	3	1.7	4 (27%)	None
Response to systemic NSAID (in idiopathic dacryoadenitis)	3	1.7	4 (29%)	None
(Epi-)scleritis overlying the involved structure	3	1	12 (44%)	Neutral
Normal adjacent sinus	4	1	17 (68%)	Agreement
Absence of superior ophthalmic vein enlargement	3	1	12 (44%)	Neutral
Normal size of supra- and infraorbital nerve	3	1	13 (48%)	Neutral
Lymphoid follicles with germinative centers	3	1.5	10 (37%)	None

Absence of granulomatous inflammation, necrosis, and/or vasculitis	4	1	21 (78%)	Agreement
Acini/duct destruction (in lacrimal gland)	3	1	9 (33%)	Neutral
Absence of epimyoeptithelial islands	3	0	6 (23%)	Neutral
<b>When IgG4-positive plasma cell infiltrates are present in the biopsy specimens - in a context of clinical, radiological and pathological indicators of IOI -, which threshold is still compatible with IOI: IgG4+/IgG ratio is ≤40%, or</b>  ≤10/hpf ≤30/hpf ≤50/hpf ≤100/hpf	<b>Lacrimal gland</b>		<b>Orbital fat/ muscle</b>	
	36.50/hpf (weighted sum)		27.22/hpf (weighted sum)	

\*a positive responder implies a 4 or 5 Likert-rating

IQR = interquartile range; IOI : idiopathic orbital inflammation, MRI = magnetic resonance imaging; CT = computerized tomography; IgG4 = immunoglobulin G4; hpf = high-power field



**eFigure.** The Initial Diagnostic Steps (With or Without Serology) Among Experts (N = 27), for Suspected IOI in the Lacrimal Gland, Muscle, or Orbital Fat

The question comprised: If idiopathic dacryoadenitis/idiopathic orbital fat inflammation\*/idiopathic orbital myositis is suspected based on the clinical and radiological context, which of the following are your initial diagnostic step(s):

Tissue biopsy\*

Corticosteroid trial

NSAID trial

Inflammatory and autoimmune serology

NSAID = nonsteroidal anti-inflammatory drugs. \*No biopsy for lesions in the orbital apex or on the optic nerve.