

# Elargissement du nerf infra orbitaire dans les orbitopathies à IgG4 : un signe spécifique

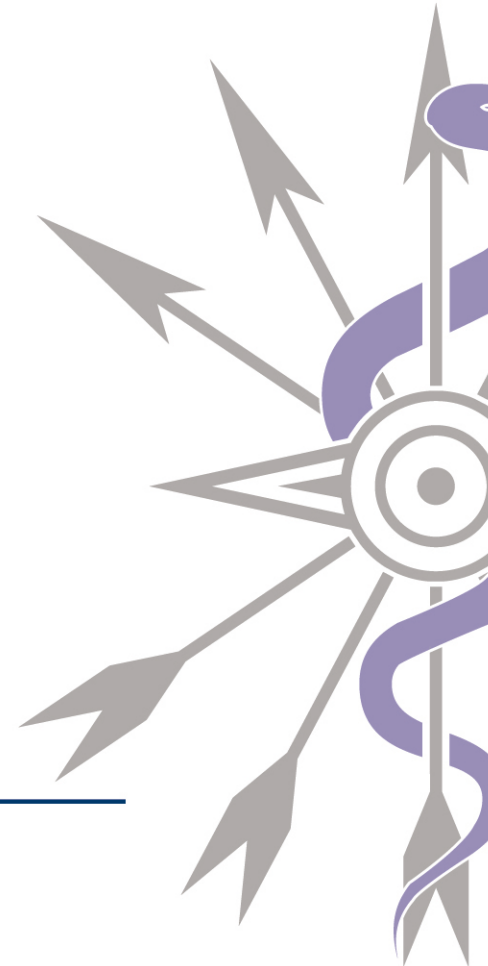


Fondation Ophtalmologique  
Adolphe de Rothschild



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Augustin Lecler  
Jérémy Bensoussan





# Conflits d'intérêt

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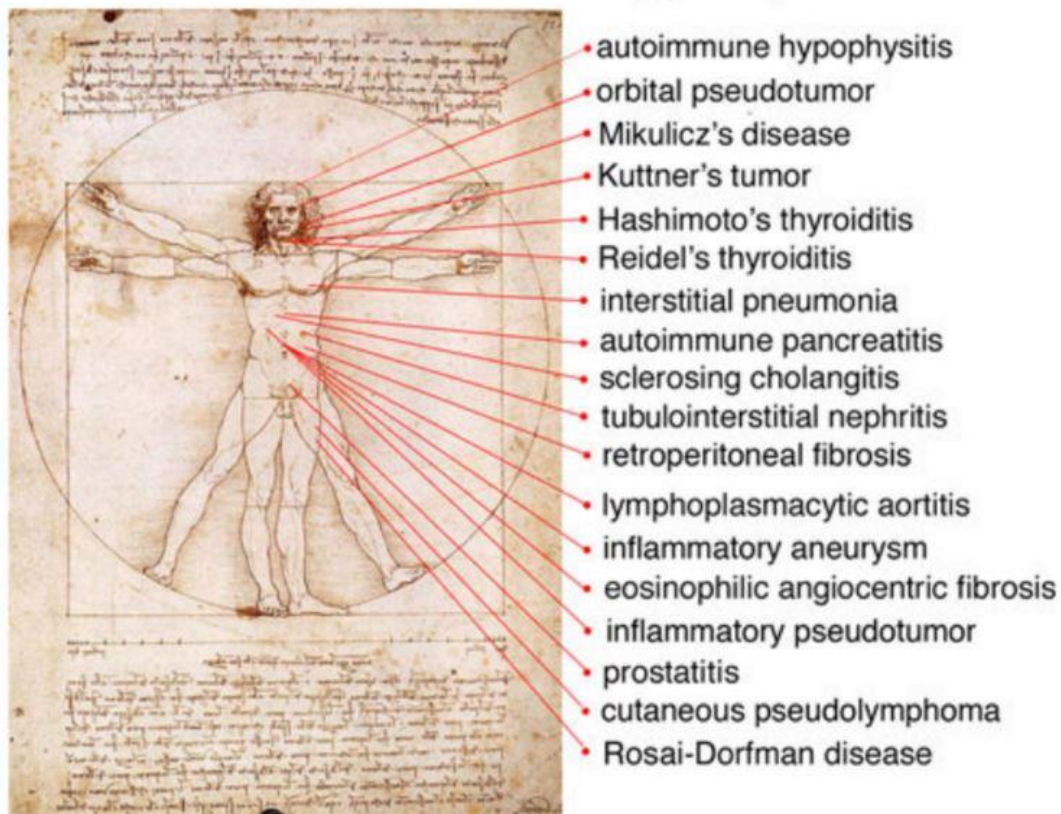
- Aucun



# IgG4

- Maladie multi systémique

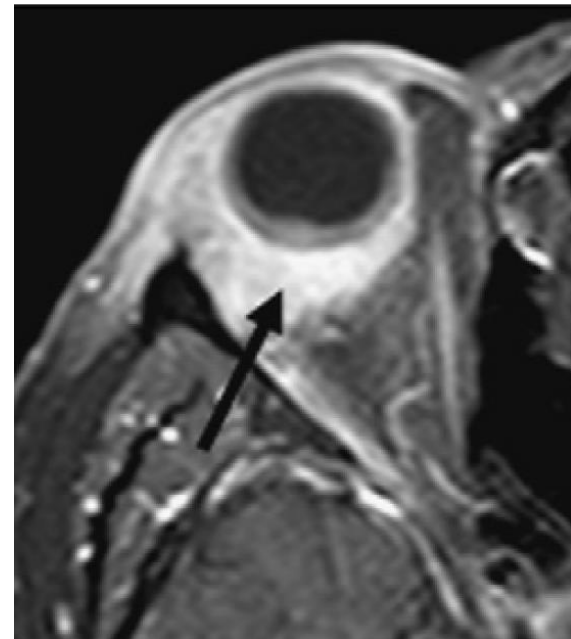
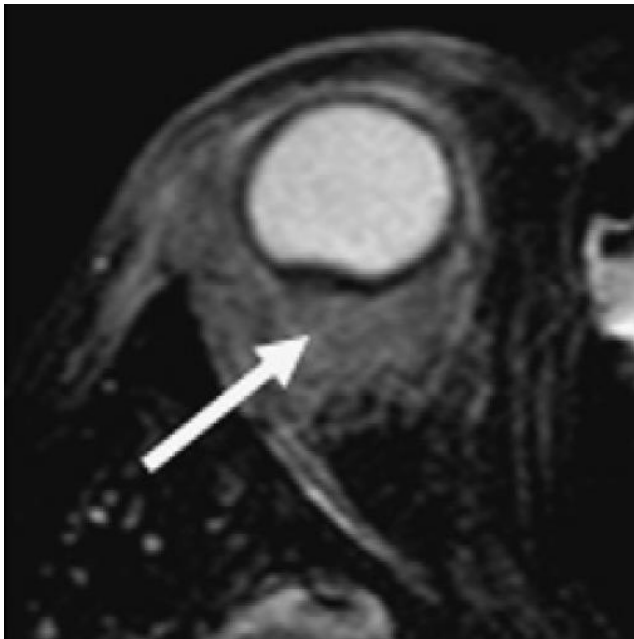
IgG4-Related Disease (IgG4RD)





# Inflammation orbitaire IgG4 (IO-IgG4)

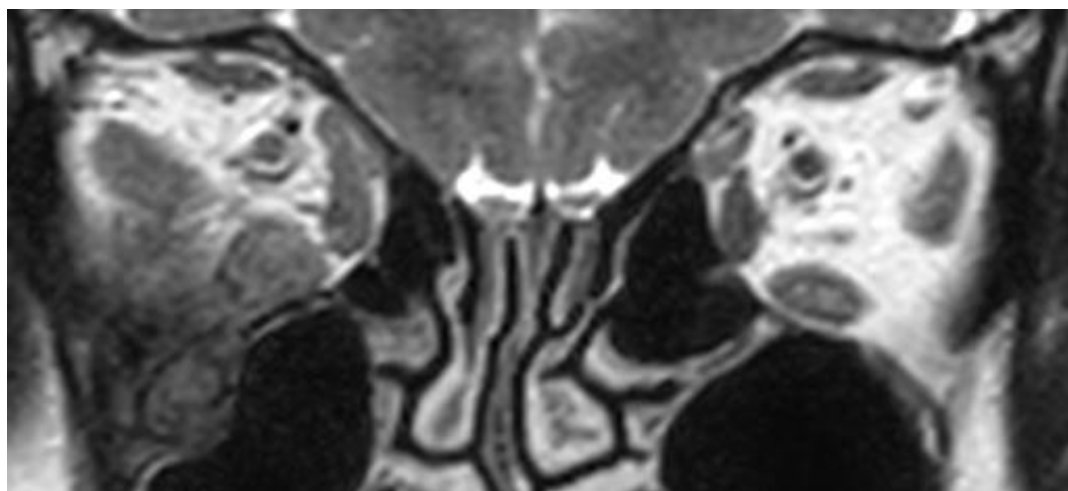
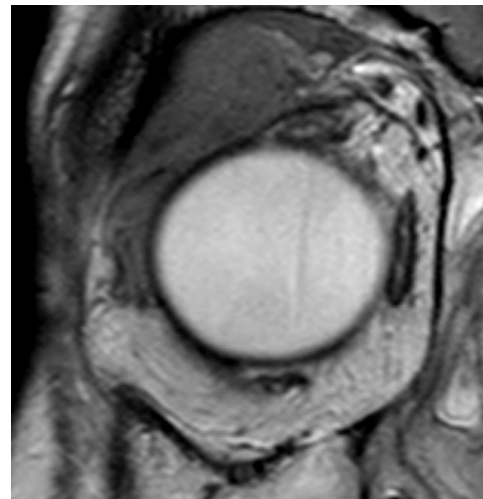
- 4-34% des IgG4
- 2<sup>nd</sup> site le plus fréquent de la région ORL
- 6%-40% des inflammations orbitaires “idiopathiques”





# IO-IgG4

- Dacryoadénite
- Myosite
- Inflammation orbitaire
- Nerf trijumeau (ENIO)

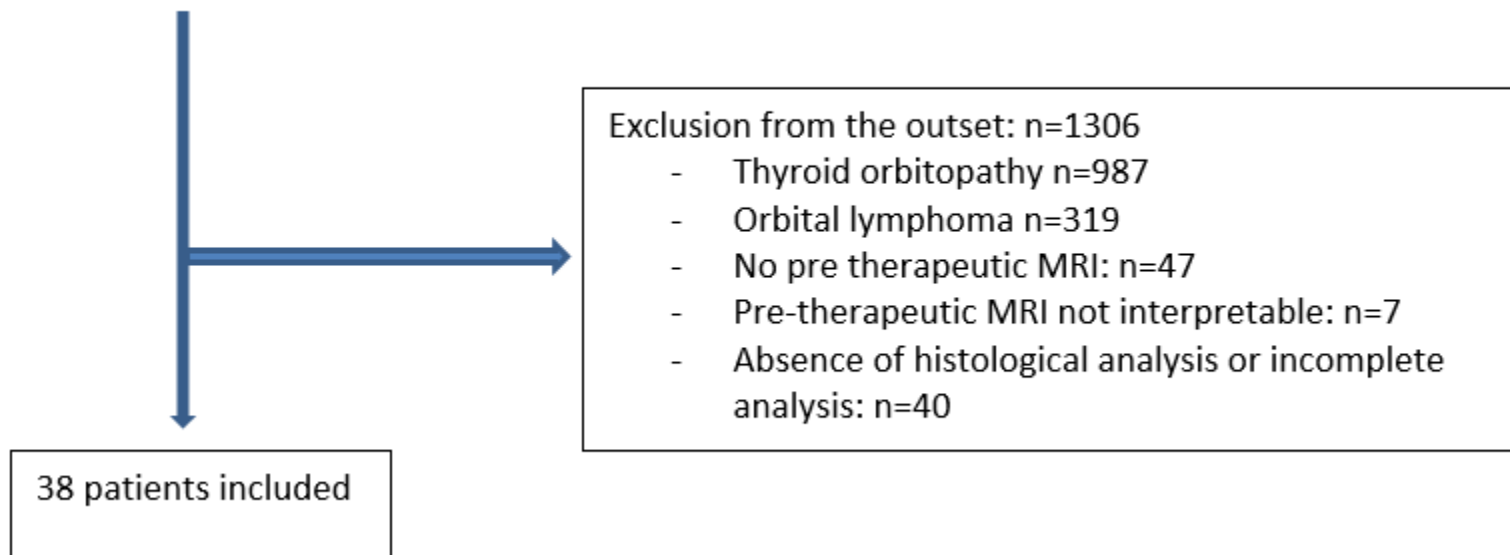




# Design de l'étude

- Retrospective, centre tertiaire
- Janvier 2006 - Avril 2015
- Deux lecteurs: junior / senior

Between January 2006 and April 2015 : n=1438 patients







# Design de l'étude

- Deux groupes

- IO-IgG4

- Non IgG4

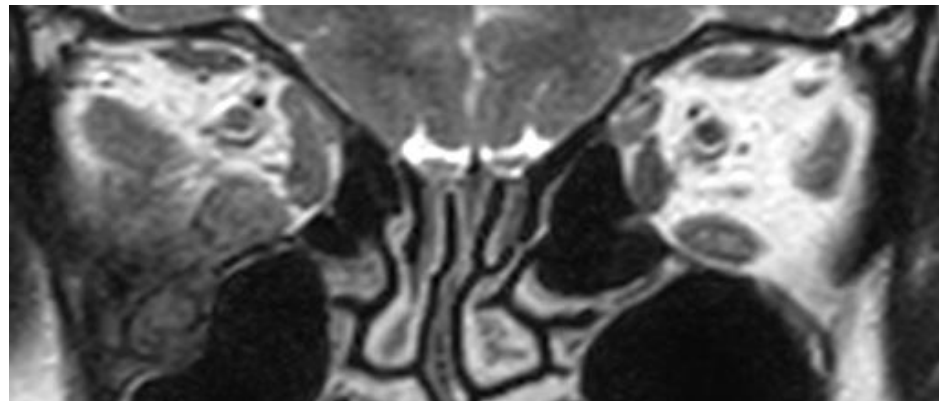
- 1 Clinical examination reveals characteristic diffuse/ localized swelling or masses in single or multiple organs

- 2 Hematological examination shows elevated serum IgG4 concentrations ( $\geq 135$  mg/dL)

- 3 Histopathological examination shows marked lymphocyte and plasmacyte infiltration and fibrosis

Infiltration of IgG4+ plasma cells: ratio of IgG4+ IgG+ cells  $>40\%$  and 10 IgG4+ plasma cells/ high power field

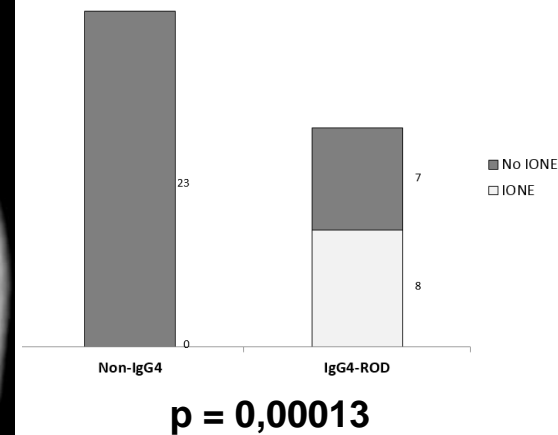
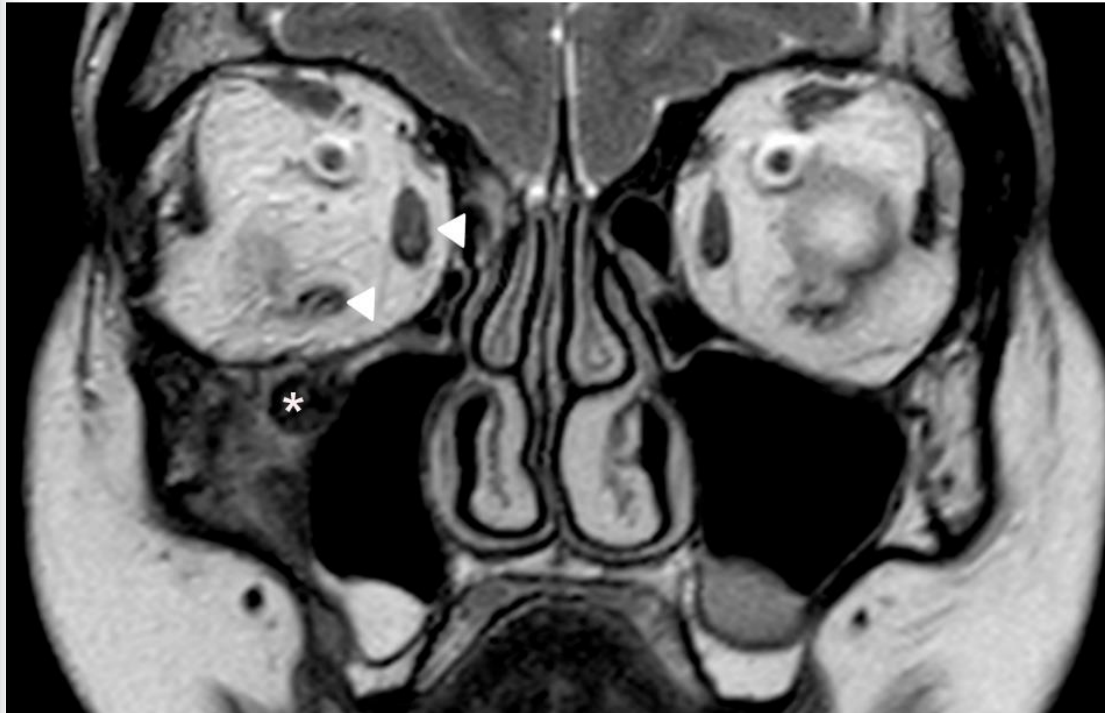
- Critère de jugement principal : ENIO





# Résultats : ENIO

- Sensibilité 53%, spécificité 100%, VPP 100%, VPN 77%

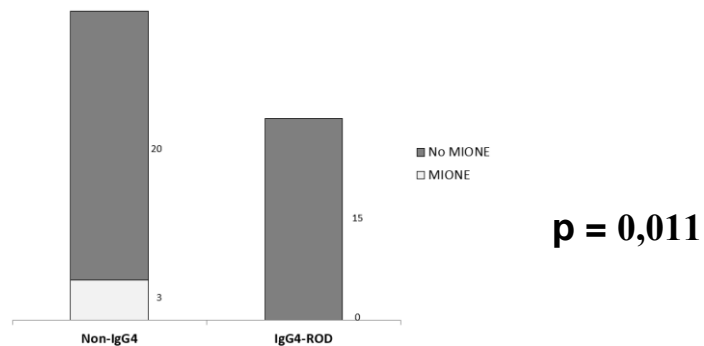
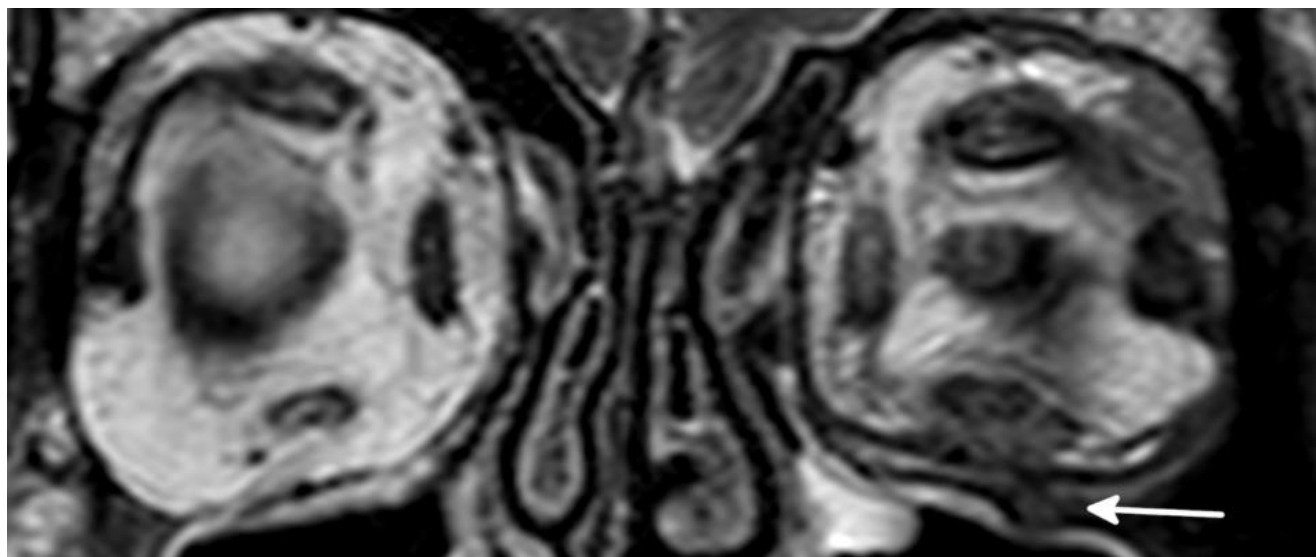






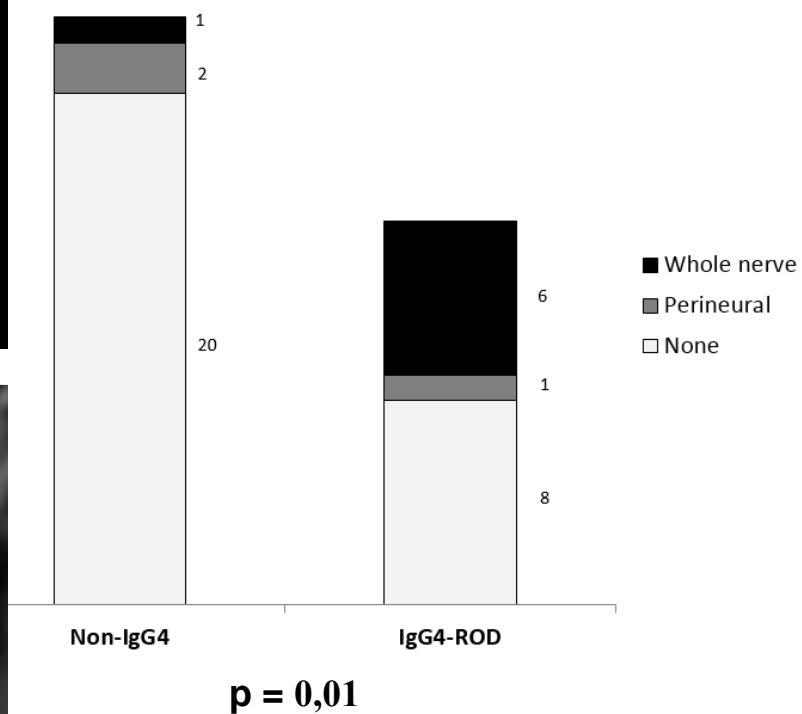
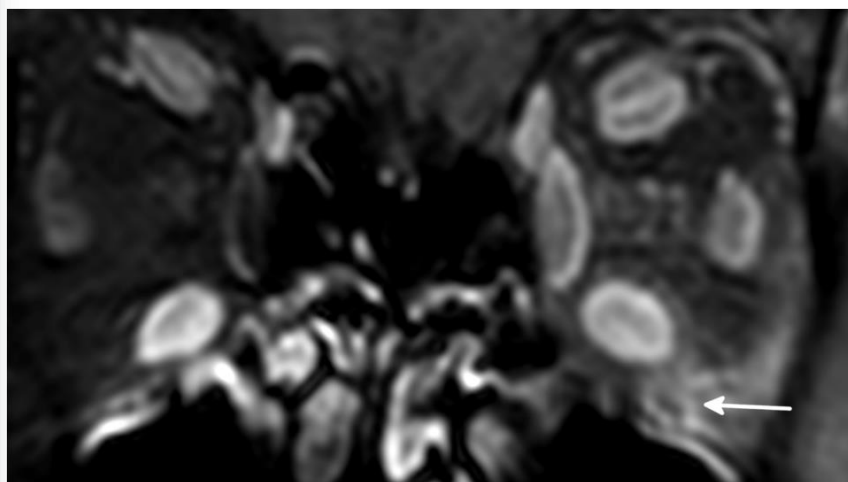
# Résultats : ENIO modéré

- ENIO modéré (plus petit que nerf optique)



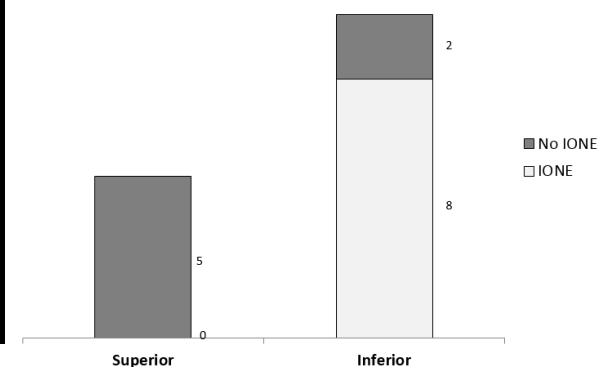
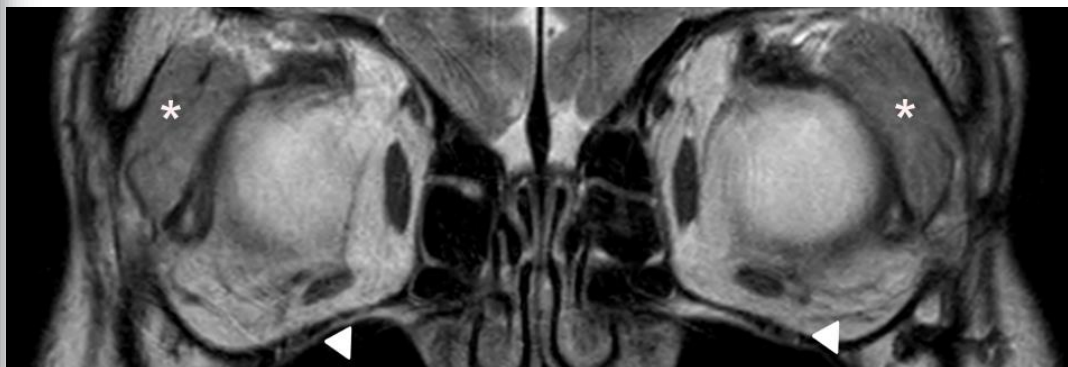


# Résultats : prise de contraste

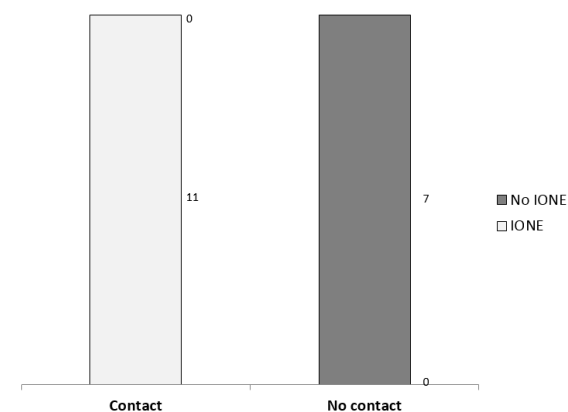
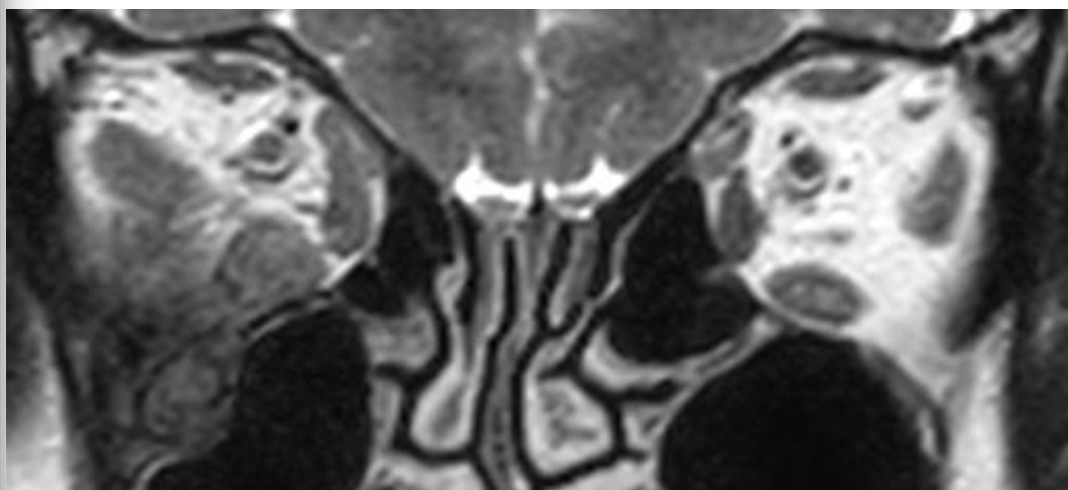




# Résultats : Contact



$p < 0.001$



$p < 0.00001$



# Discussion

- Littérature

	IgG4-ROD	IONE	Trigeminal involvement	Mean age	Sex	Bilaterality
<b>Ohshima et al 2012</b>	n = 16	40.9%	NA	NA	NA	NA
<b>Sogabe et al 2014</b>	n = 65	32%	38,5%	59,4	31M/34W	53%
<b>Takano et al 2014</b>	n = 68	29.4%	NA	58,8	31M/37W	NA
<b>Inoue et al 2012</b>	n = 6	23.8%	42.9%	NA	NA	NA
<b>Katsura et al 2012</b>	n = 17	NA	24%	66,1	15M/2W	NA
<b>Tiegs-Heiden et al 2014</b>	n = 27	30%	NA	51	10M/17W	88%
<b>Wu et al 2015 (Meta-analysis)</b>	n = 172	NA	23-39%	57	90M/82W	68%
<b>Our series</b>	n = 15	53%	87%	47,6	6M/9W	13%



# Discussion

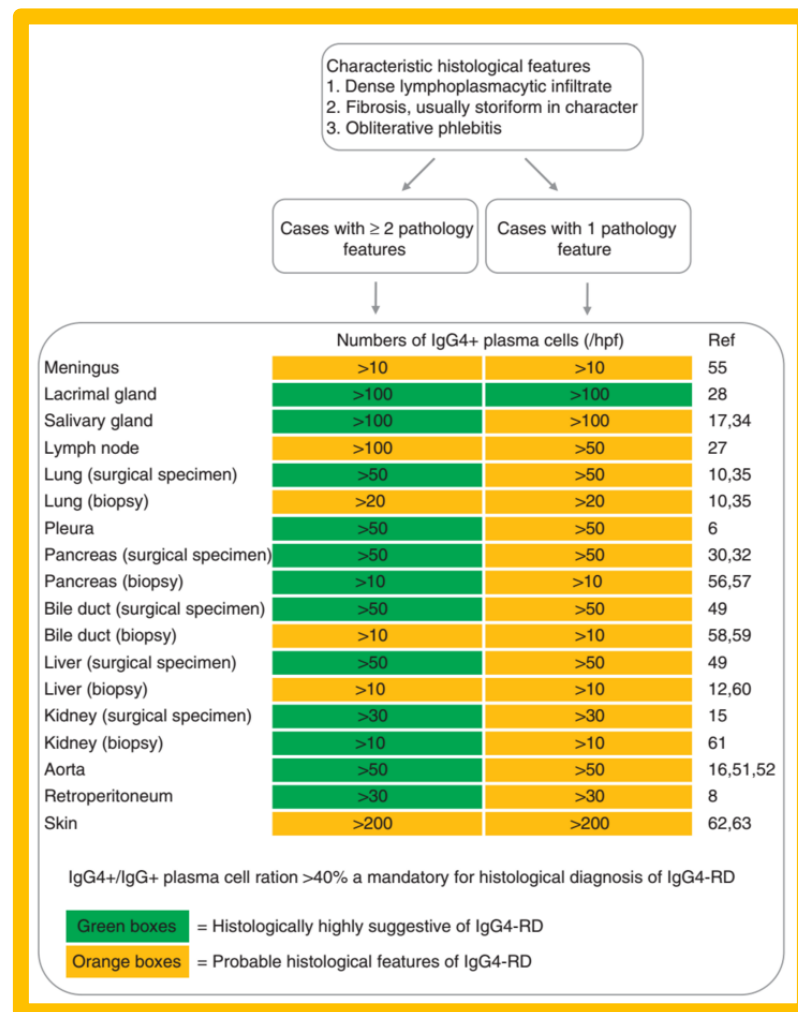
## • Critères diagnostiques

- 1 Clinical examination reveals characteristic diffuse/localized swelling or masses in single or multiple organs
- 2 Hematological examination shows elevated serum IgG4 concentrations ( $\geq 135$  mg/dL)
- 3 Histopathological examination shows marked lymphocyte and plasmacyte infiltration and fibrosis  
Infiltration of IgG4+ plasma cells: ratio of IgG4+ IgG+ cells  $>40\%$  and 10 IgG4+ plasma cells/high power field

**Table 2** Diagnostic criteria for IgG4 related ophthalmic disease, 2014

- (1) Imaging studies show enlargement of the lacrimal gland, trigeminal nerve, or extraocular muscle as well as masses, enlargement, or hypertrophic lesions in various ophthalmic tissues
- (2) Histopathologic examination shows marked lymphocyte and plasmacyte infiltration, and sometimes fibrosis. A germinal center is frequently observed. IgG4+ plasmacytes are found and satisfy the following criteria: ratio of IgG4+ cells to IgG+ cells of 40 % or above, or more than 50 IgG4+ cells per high-power field ( $\times 400$ )
- (3) Blood test shows elevated serum IgG4 ( $\geq 135$  mg/dl)

Diagnosis is classified as “definitive” when (1), (2), and (3) are satisfied; “probable” when (1) and (2) are satisfied; and “possible” when (1) and (3) are satisfied

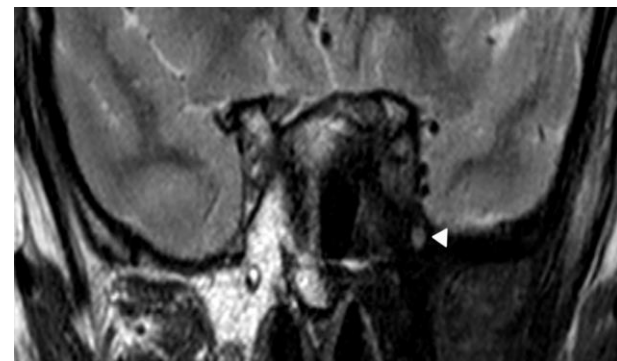
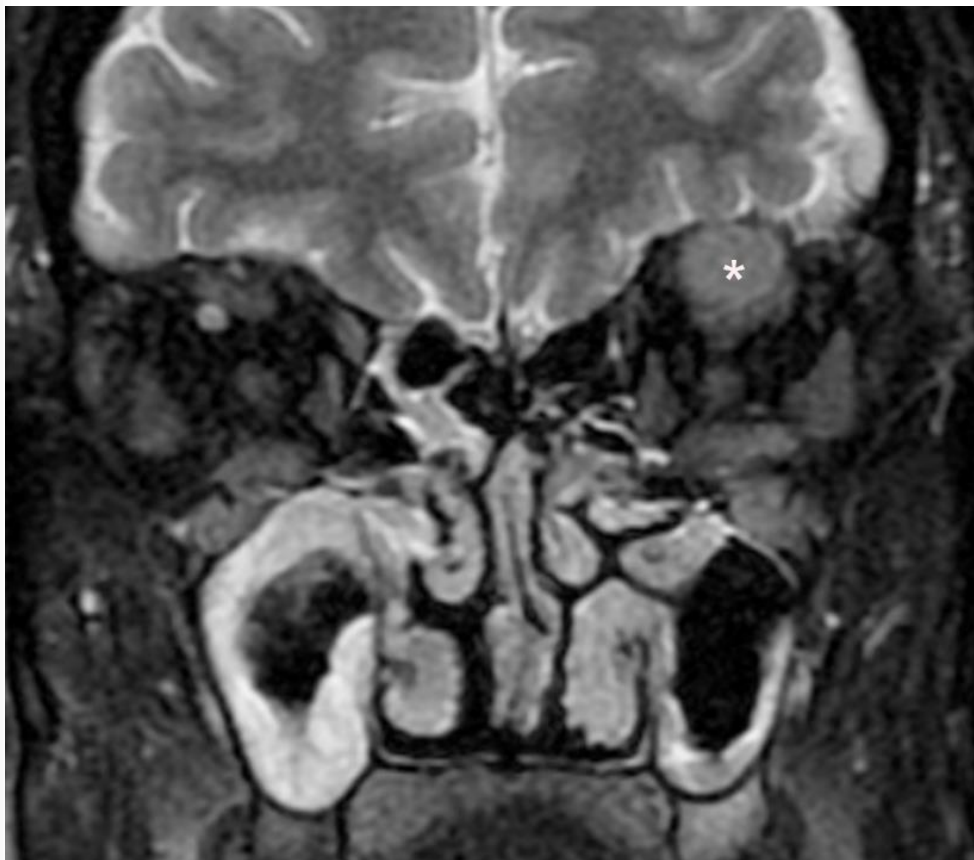






# Discussion

- Extension périneurale







# Limites

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- Retrospectif
- Faible nombre de patients
- Fort taux d'exclusion
- IgG4 sériques non disponibles



# Conclusion

- ENIO : hautement spécifique
- Signe clé pour le diagnostic

