



Ophthalmic Oncology Service
Memorial Sloan Kettering Cancer Center

Spectrum of clinical and subclinical choroidal abnormalities in patients with histiocytosis

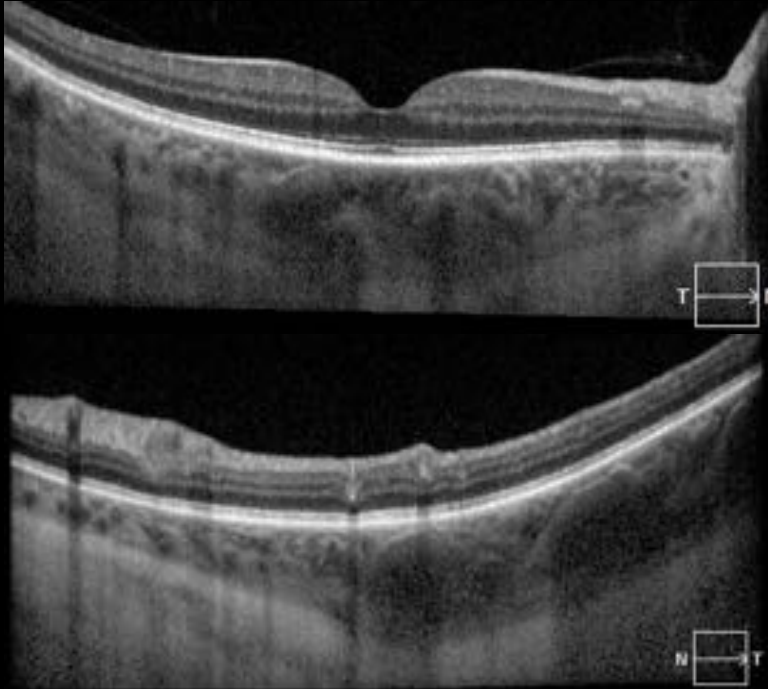
Jasmine H. Francis MD, FACS

Julia Canestraro OD FAAO, Rampal Raajit MD, David H Abramson MD FACS,
Eli L Diamond MD

Retina Society Meeting 2020

No Financial Interests or Relationships
Off label Use Drugs

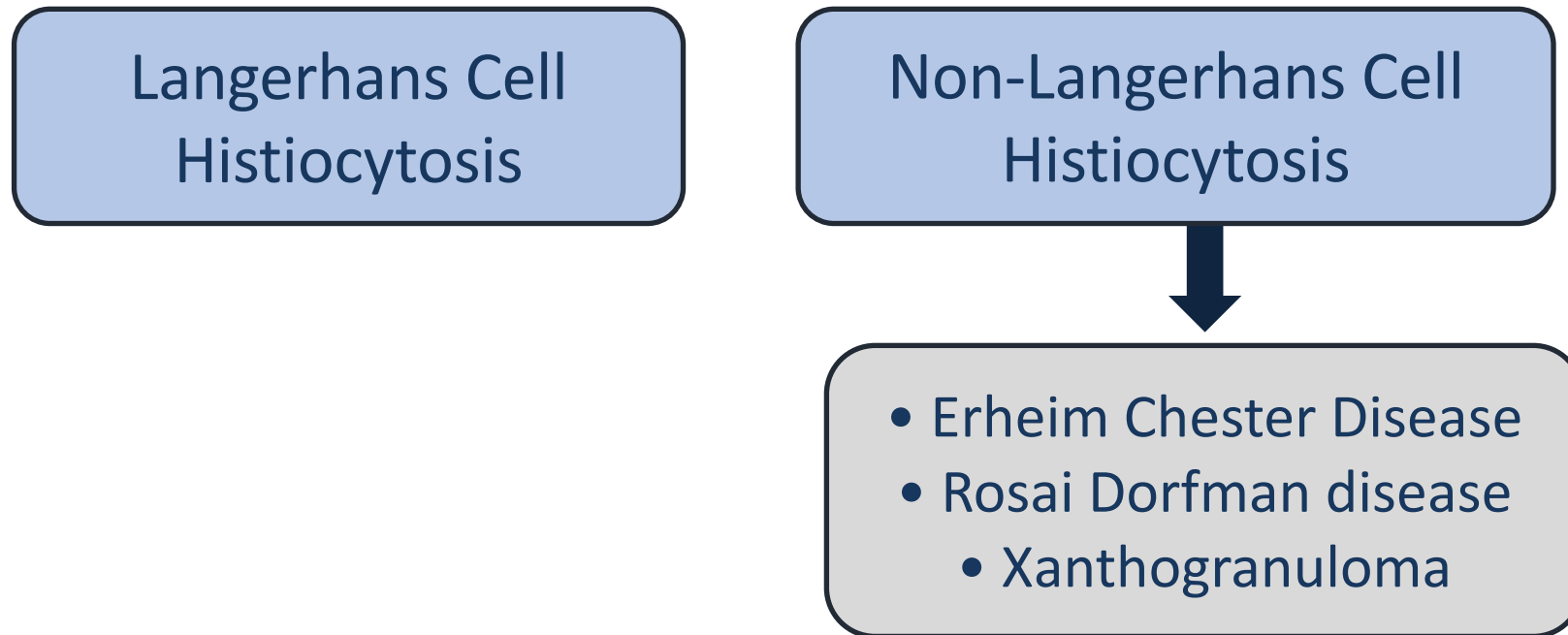
Conclusion/Summary



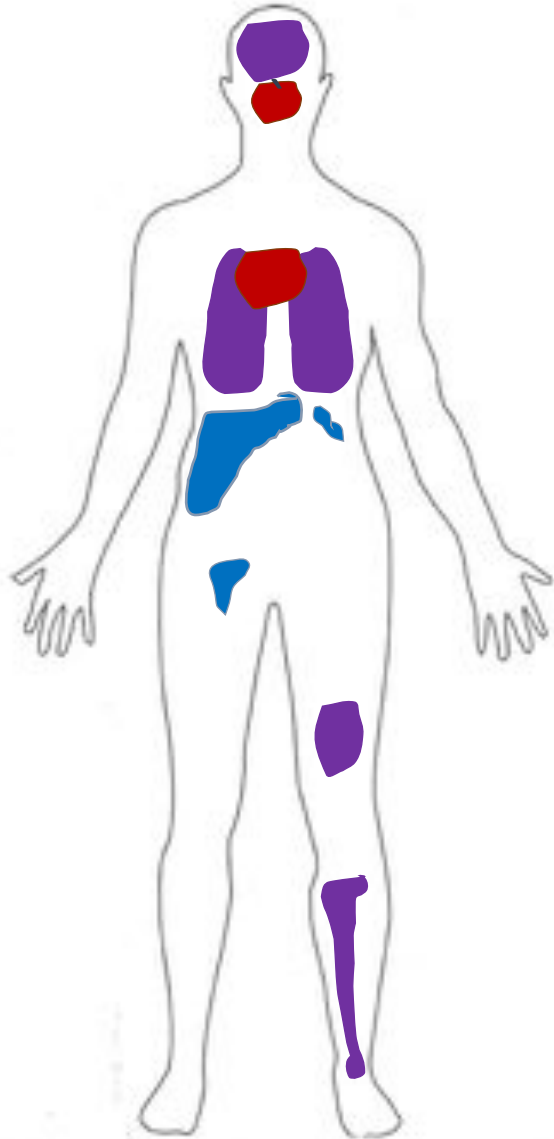
- 14% of non-LCH histiocytosis patients have clinically evident infiltration of the choroid
- However, the vast majority of these patients (80%), have statistically significant increase in SFCT compared to age- and gender-matched controls
- With systemic treatment, SFCT decreases
- Choroid morphology:
 - outer retinal infiltrative densities (40%)
 - enlarged Haller's layer w/ inner ret. distortion (60%)
- There is no association with CNS nor osseous disease

Histiocytic Disorders: Introduction

- Clonal, hematopoietic disorders
- Accumulations of activated histiocytes in affected tissues



Histiocytic Disorders: multisystem disease



Langerhans Cell

CNS (6%)

Hypothalamic-pituitary axis

Lung (10%)

Spleen (13%)

Liver (16%)

Lymph nodes (19%)

Skin (39%)

Bone (77%)

Non-LCH (ECD)

CNS (40%)

Lung (46%)

Orbit (22%)

Maxillary sinus (59%)

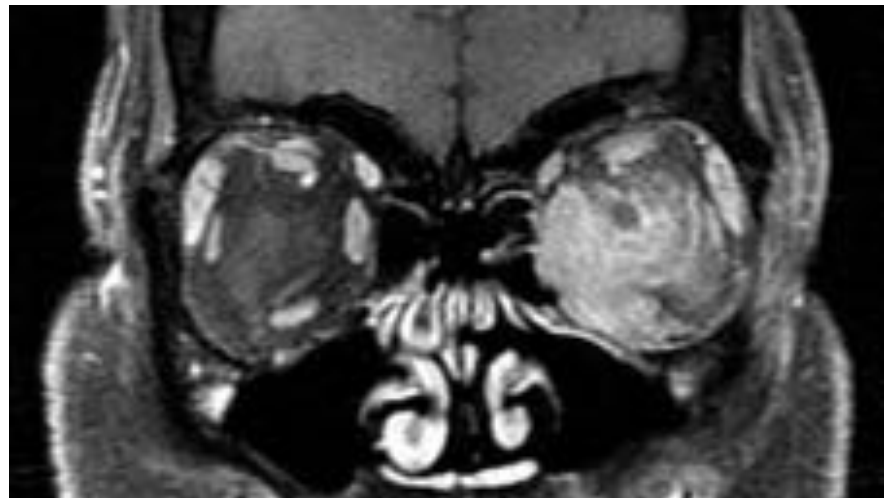
Heart (57%)

Skin (27%)

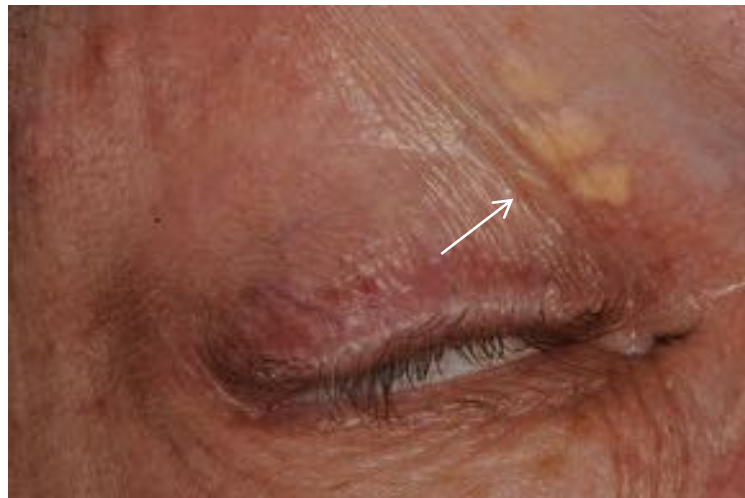
Bone (95%)

Histiocytic Disorders: Ocular manifestations

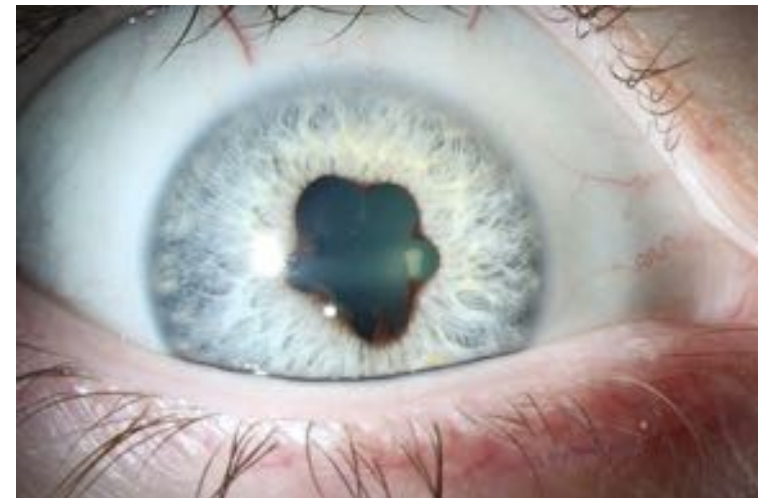
Orbit



Eyelid



Other...



Histiocytic Disorders: Published Choroid

Choroidal Langerhans' cell histiocytosis

Acta Ophthalmol Scand, 2000
In Taek Kim and Sang Min Lee

Kim & Kim, Acta Ophthalmol Scand, 2000

Presumed Choroidal Langerhans Cell Histiocytosis Following a Previously Resected Solitary Central Nervous System Lesion in an Adult

Niall Patton, MRCOphth
Tze Lai, MBBS(Hons)
Peter Robbins, FRCPA
David Holthouse, MBBS(Hons)
Chris Barry, MMedSci
Ian Constable, FRACO, FRACS
Arch Ophthalmol, 2006

Ophthalmol, 2006

Choroidal Involvement in Erdheim-Chester Disease

Amro Abdellatif, MBBS; Craig M. Mason, MD;
Steven R. Ytterberg, MD; Stephen A. Boorjian, MD;
Diva R. Salomão, MD; Jose Pulido, MD, MS, MPH

1

Abdellatif et al. Ophthalmic Surg Lasers Imaging Retina, 2015

Graefes Arch Clin Exp Ophthalmol (2013) 251:295–299
DOI 10.1007/s00417-012-2175-5

PATHOLOGY

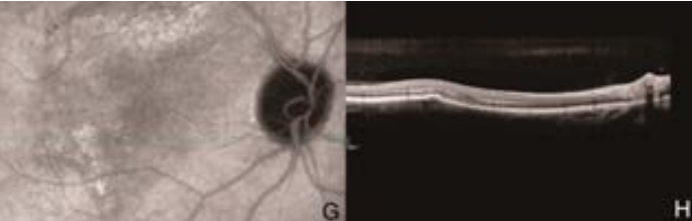
Rosai–Dorfman disease presenting as choroidal melanoma: a case report and review of the literature

Tersia L. Vermeulen · Timothy W. Isaacs ·
Dominic Spagnolo · Benhur Amanuel

Vermeulen et al, Graefes Arch Clin Exp Ophthalmol, 2013

ROSAI–DORFMAN DISEASE DIAGNOSED BECAUSE OF BILATERAL CHOROIDAL MASSES

Yoreh Barak, MD, Melissa G. Tong, MD, Todd J. Purkiss, MD, Shlomit Schaal, MD, PhD
Retin Cases Brief Rep, 2012



Barak et al, Retin Cases Brief Rep, 2012

Graefes Arch Clin Exp Ophthalmol (2015) 253:1819–1820
DOI 10.1007/s00417-015-2987-1

LETTER TO THE EDITOR

Choroidal involvement in Rosai–Dorfman syndrome may be depicted and followed using enhanced depth imaging optical coherence tomography (EDI-OCT)

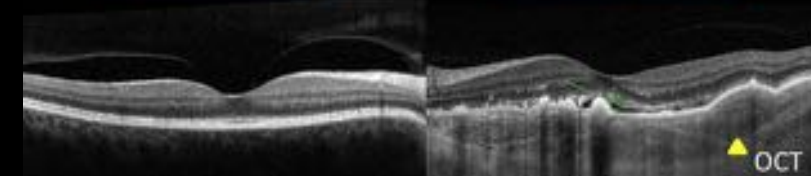
Janelle Fassbender¹ · Shlomit Schaal¹

Fassbender et al, Graefes Arch Clin Exp Ophthalmol, 2015



Three Cases of Erdheim-Chester Disease With Intraocular Manifestations: Imaging and Histopathology Findings of a Rare Entity

ANNA C.S. TAN, SUZANNEYZER, NEAL ATEBARA, BRIAN P. MARR, ROBERT M. VERDIJK, VIRGIL A.S.H. DALM, K. BAILEY FREUND, LAWRENCE YANNUZZI, J. ANTONIOLOTTI



Tan et al. Am J Ophthalmol, 2017

Study Question

In histiocytosis:

1. Does the OCT of the choroid have characteristic findings?
2. Does this change with treatment?

Histiocytosis Service
Memorial Sloan Kettering

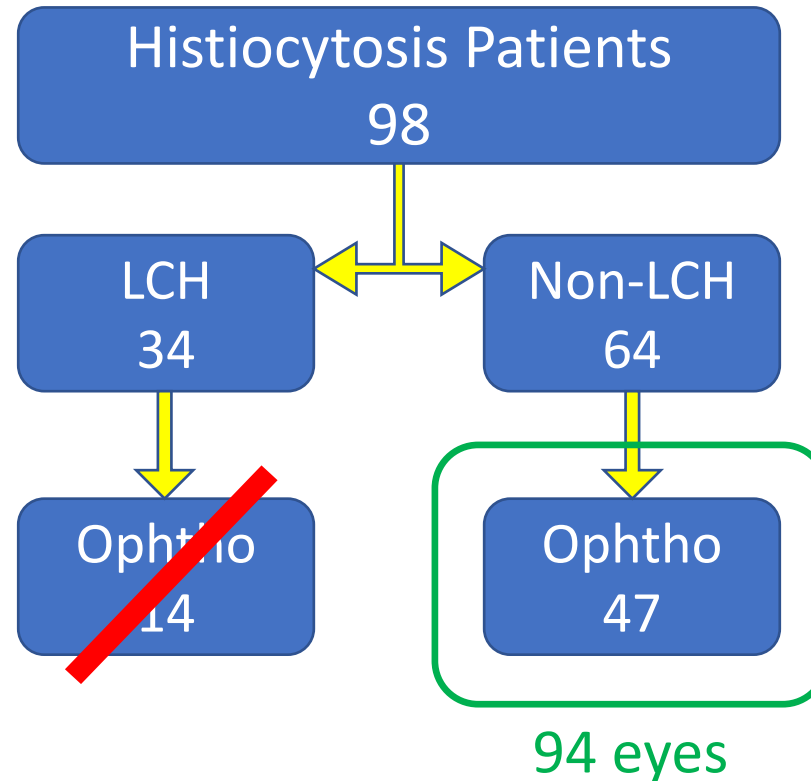
<https://www.mskcc.org/cancer-care/types/histiocytosis>



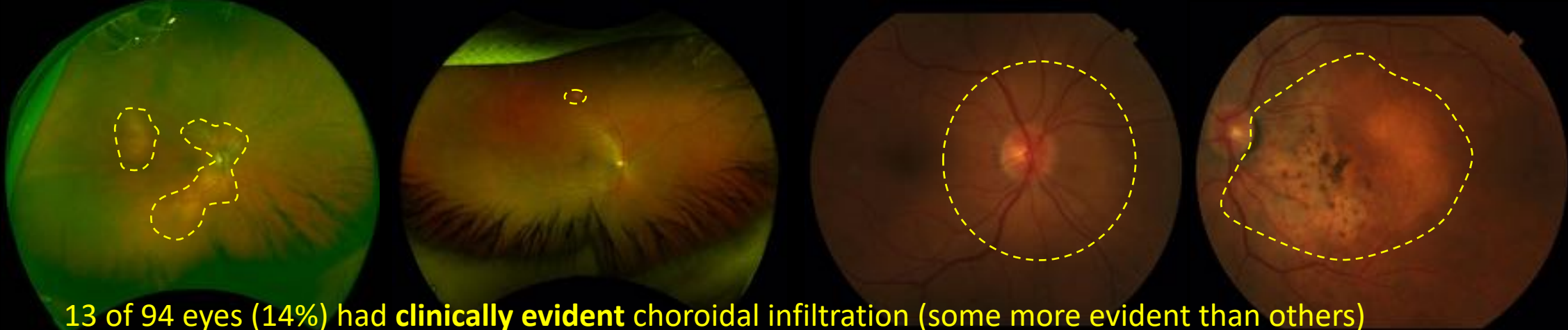
Eli Diamond MD

Methods

Patients included:



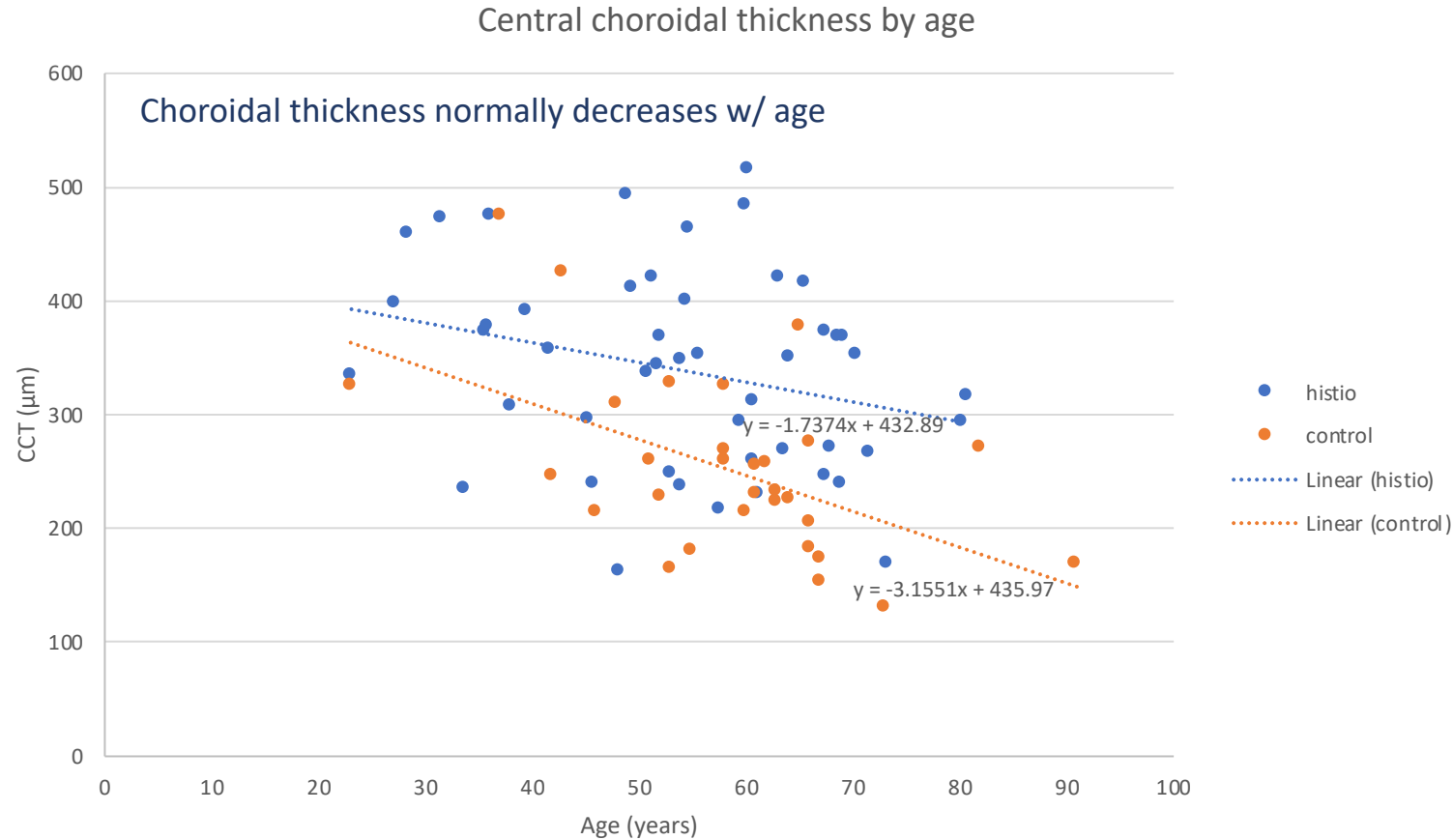
- This was compared to an age and gender matched control set of patients



13 of 94 eyes (14%) had clinically evident choroidal infiltration (some more evident than others)
In all cases, the findings are post-equatorial. In the 9 patients, 4 had bilateral findings.



Results



Mean subfoveal choroidal thickness (SFCT):

Normal controls = 250 μm
Non-LCH patients = 337 μm .

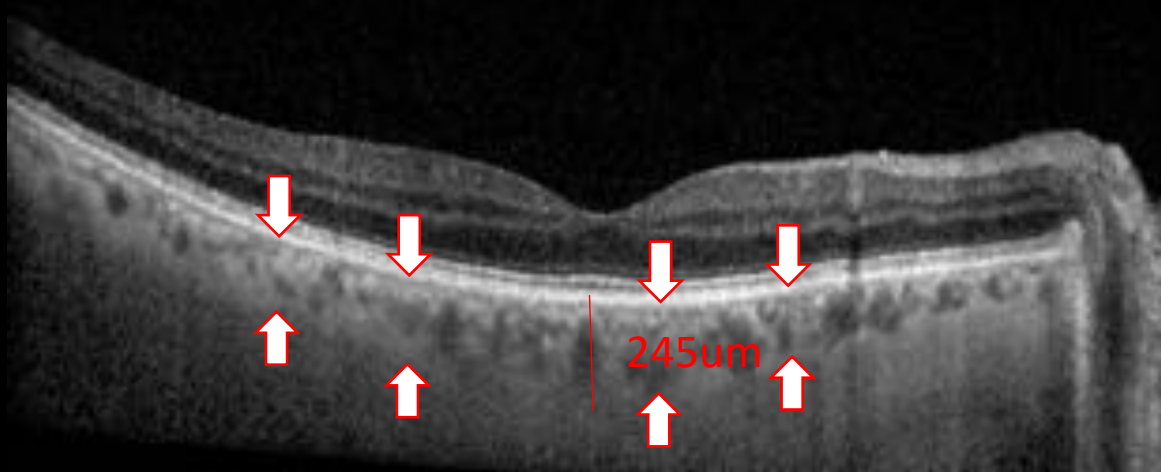
The difference, $p = 0.00008$

- Non-LCH patients had thicker SFCT, as measured by OCT

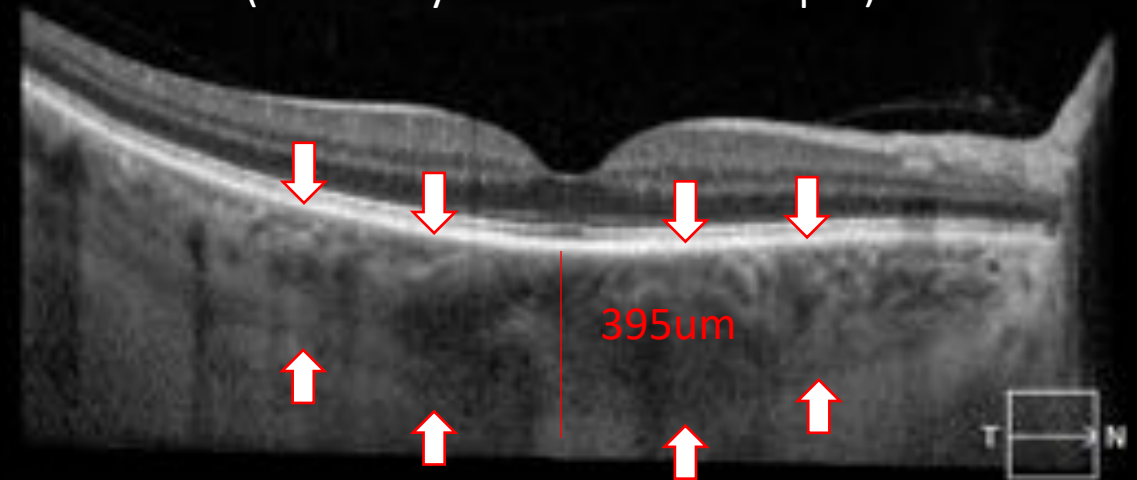
Results

Examples of choroidal thickness

NORMAL: mean 250 μ m



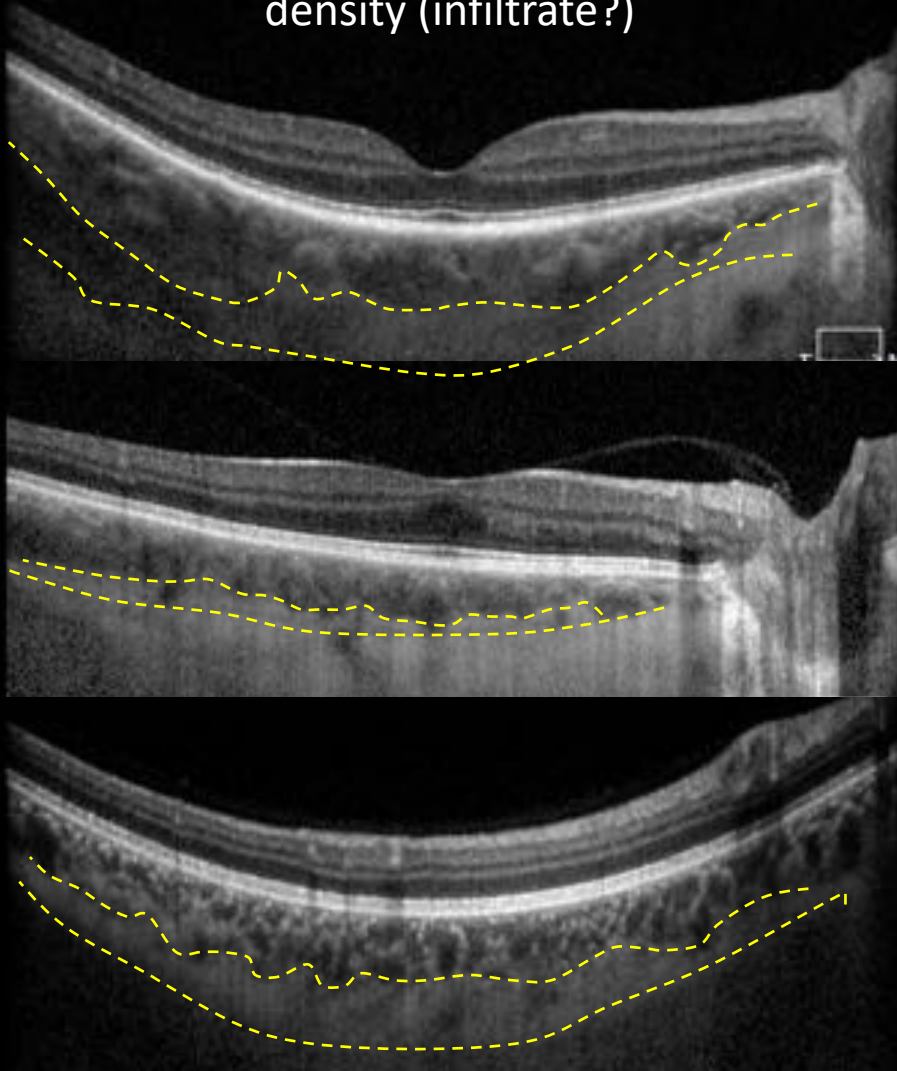
HISTIO: mean 337 μ m
(80% of eyes had SFCT > 250 μ m)



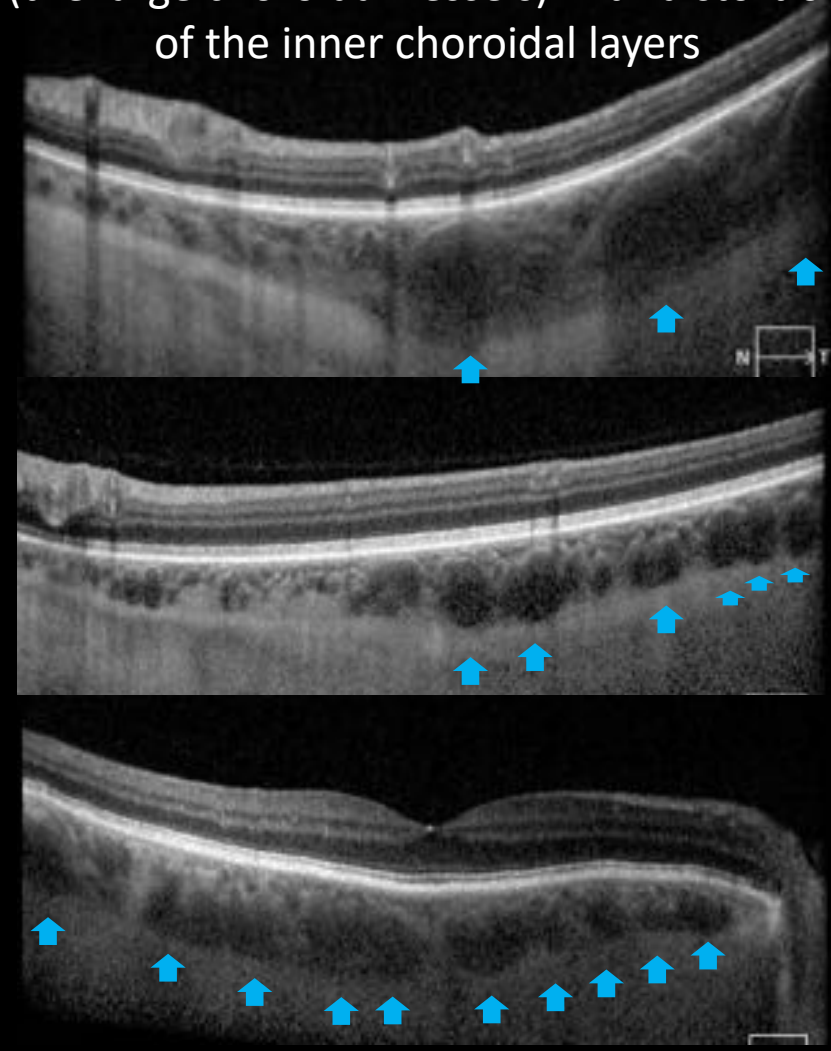
Results

Other occult choroidal findings in non-LCH eyes

40 % of eyes have an outer choroidal density (infiltrate?)

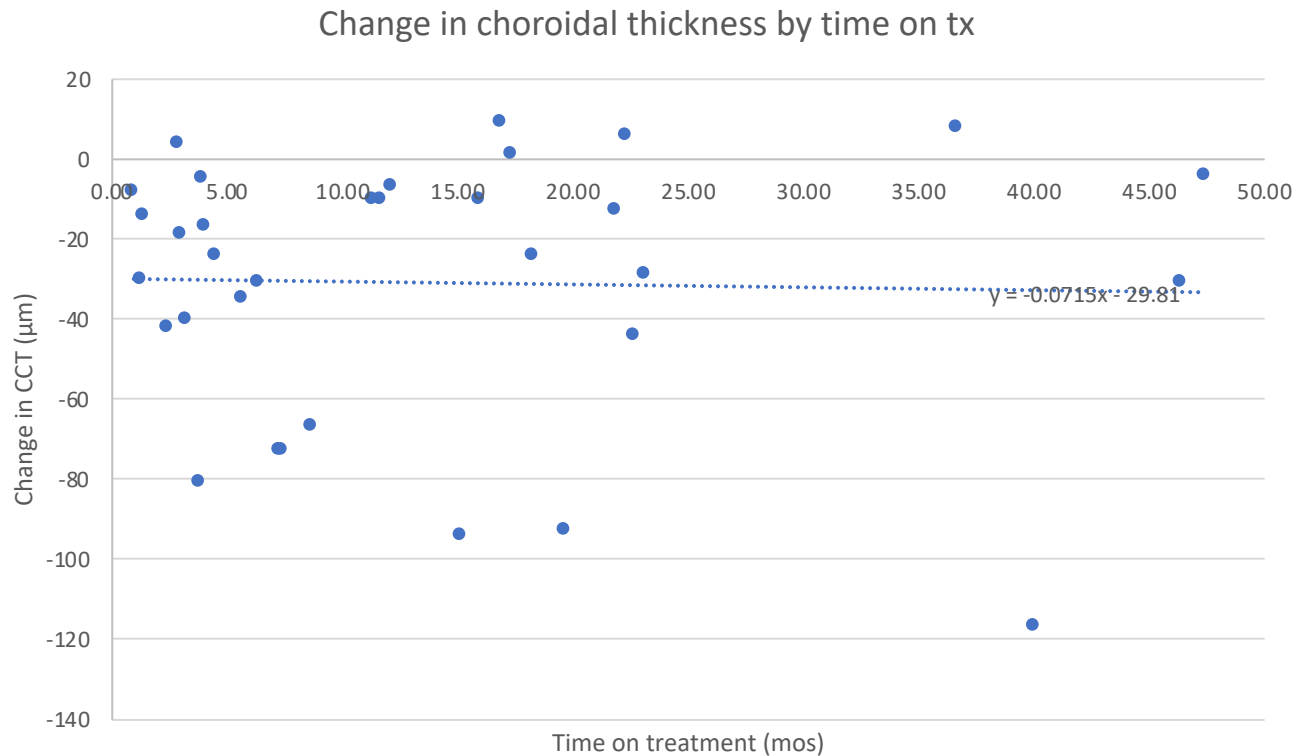


60 % of eyes have an enlarged Haller's layer (the large choroidal vessels) with distortion of the inner choroidal layers



Results

- 34 non-LCH patients (with imaging at baseline AND follow up): significant decrease in SFCT treatment on systemic treatment



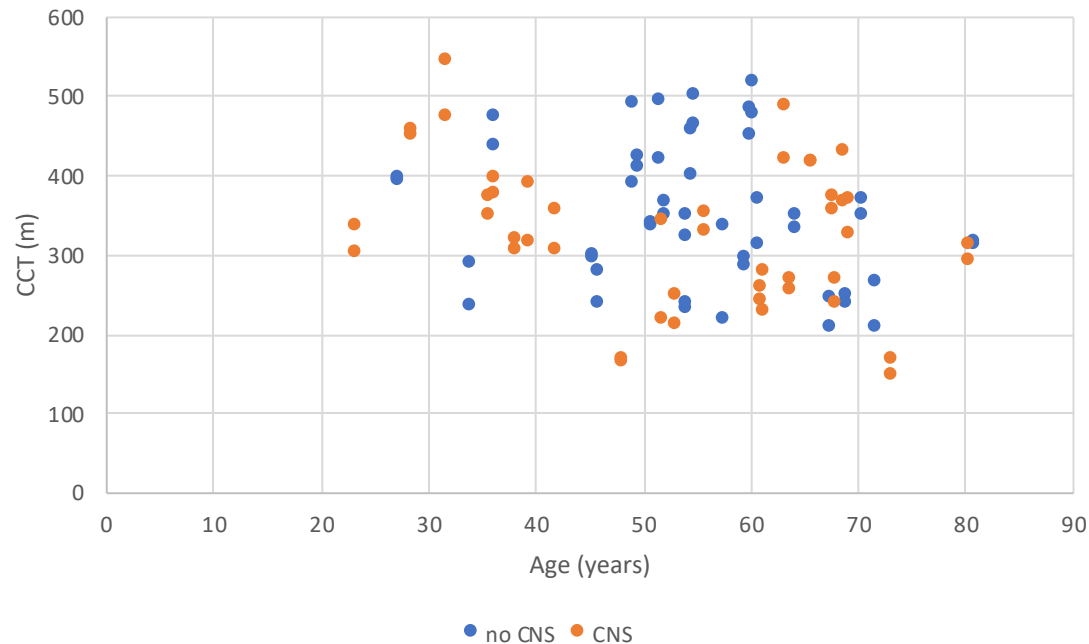
The mean change was a decrease of $31\mu\text{m}$ (p-value = 0.000005)

The length of time on treatment (and f/u exam) did not appear to influence the amount of SFCT

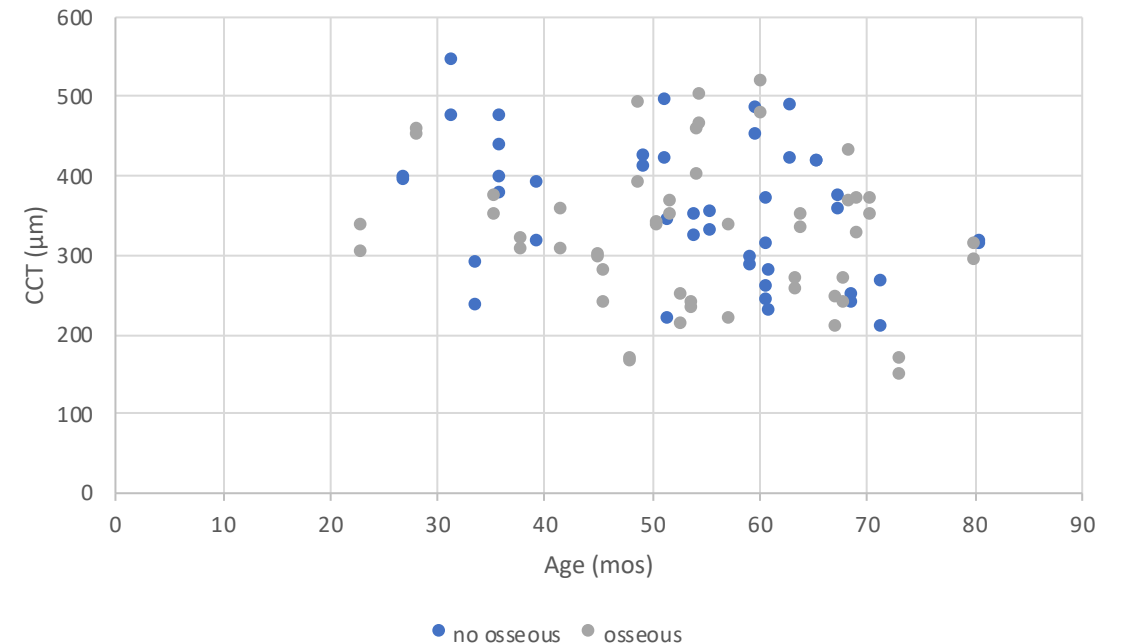
Results

Is CNS/osseous disease a marker for disease burden and would that correlate with choroidal thickness?

- No significant difference in SFCT between patients with or without CNS disease (p-value = 0.35)



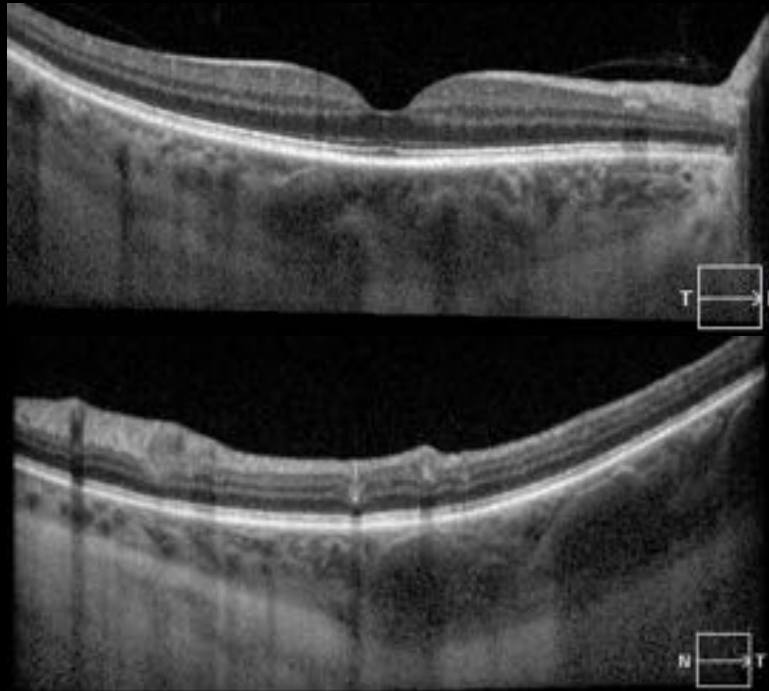
- No significant difference in SFCT between patients with or without osseous disease (p-value = 0.35)



Why is the choroid thicker in histio?

- We know the choroid is a site for accumulation of activated histiocytes (in the clinically evident cases). In the subclinical eyes, could the thicker choroid represent disease involvement?
- *For instance, lymphoma and myeloproliferative diseases can infiltrate the choroid*
- And/or could the choroid be a site where these abnormal activated histiocytes are being generated?
- We know the choroid can be a site of extramedullary hematopoiesis (both in neonates and adults- although our knowledge is limited to enucleation specimens). Therefore, presumably, the choroid could be a source of these cells

Conclusion/Summary



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