

triade capitale

cerebral vasculopathy

RED-M retinopathy, encephalopathy, deafness, microangiopathy

S SMALL

I INFARCTIONS

C COCHLEAR

R RETINAL

E ENCEPHALIC

T TISSUES

syndrome de SUSAC

SNC

SUBAIGUE, MULTIFOCALE

« MIGRAINE », SCOTOME SCINTILLANT: 3/4

PSYCHOSE AIGUE

CONFUSION, DYSARTHRIE, MUTISME

biologie méningite

LCR :

HYPERPROTEINORACHIE (0.2 - 2.8 G/L)

gammopathie oligoclonale possible

PLEIOCYTOSE (1-85, 15 éléments en moyenne)

IRM cerveau

hyperΣ T2 et T2-FLAIR

discrets et petits (1 - 7 mm)

supra- et infra-tentorie

s.blanche et/ou noyaux gris (70%)

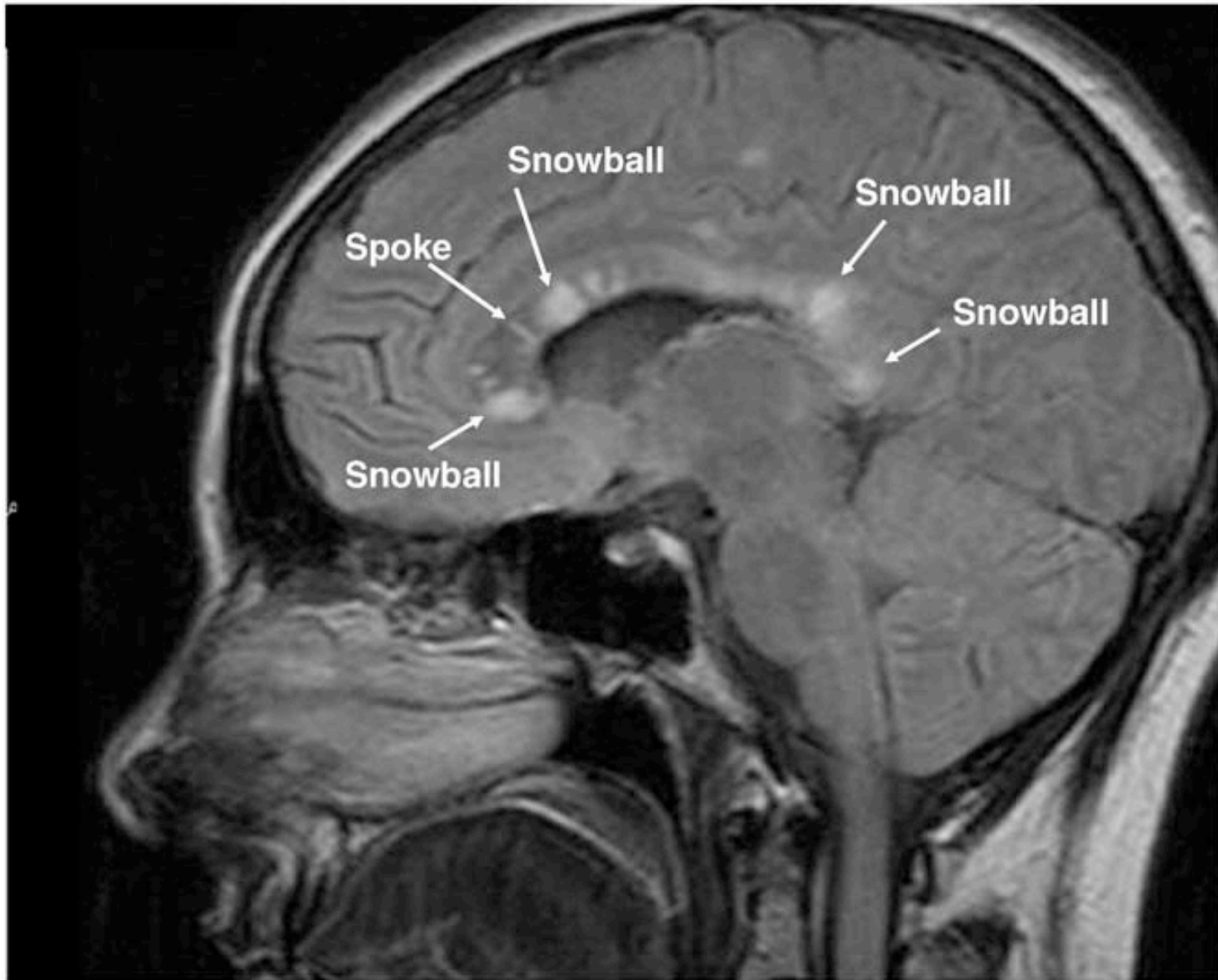
corps calleux, centre *sine qua non*
diffusion

gado + en T1 « leptoméninge »

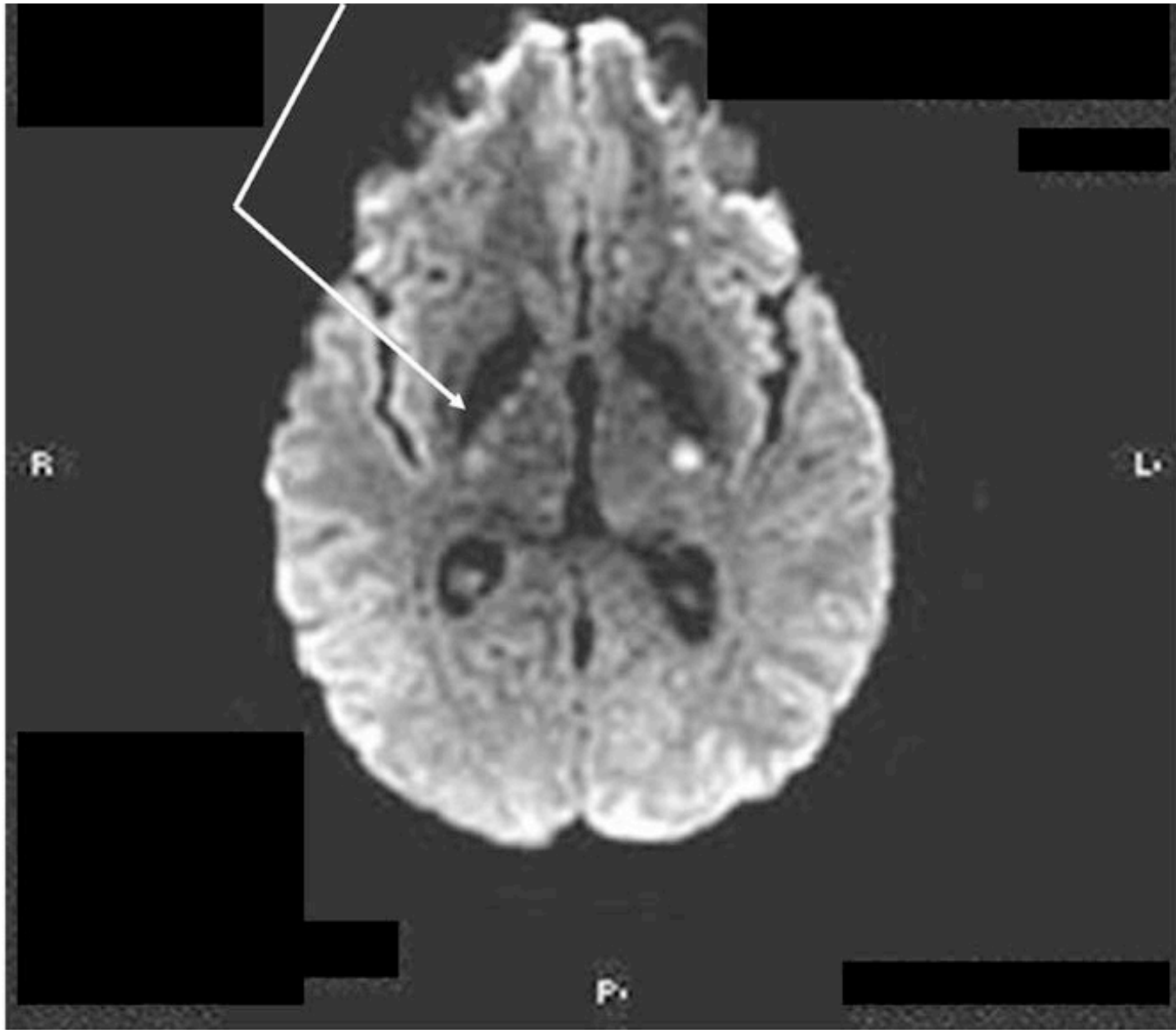
pas de // radio-clinique

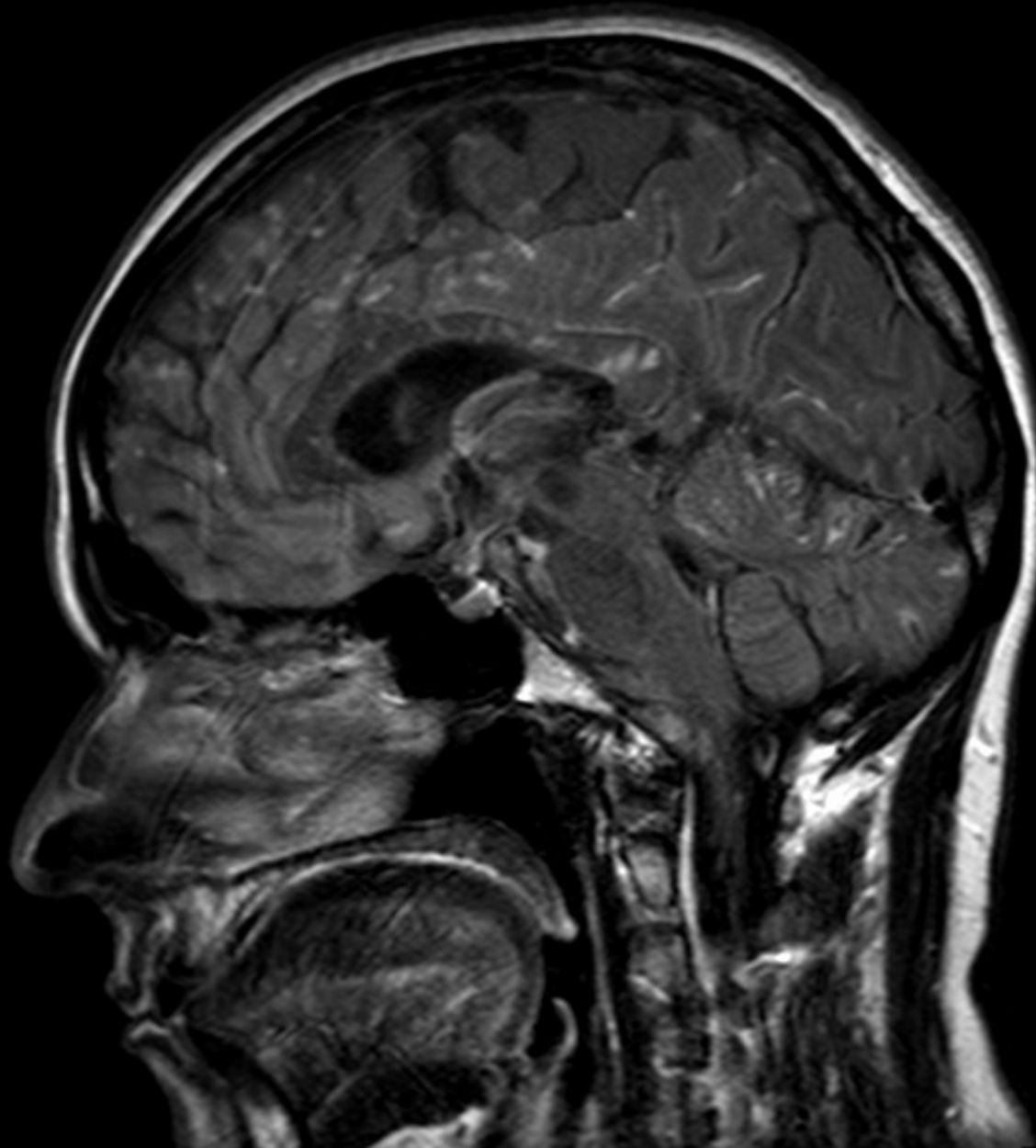
réversibilité

corps calleux : snow balls, spokes



diffusion : string of pearls





RETINE

clinique

OCCLUSION BRANCHES ACR BILATERALE

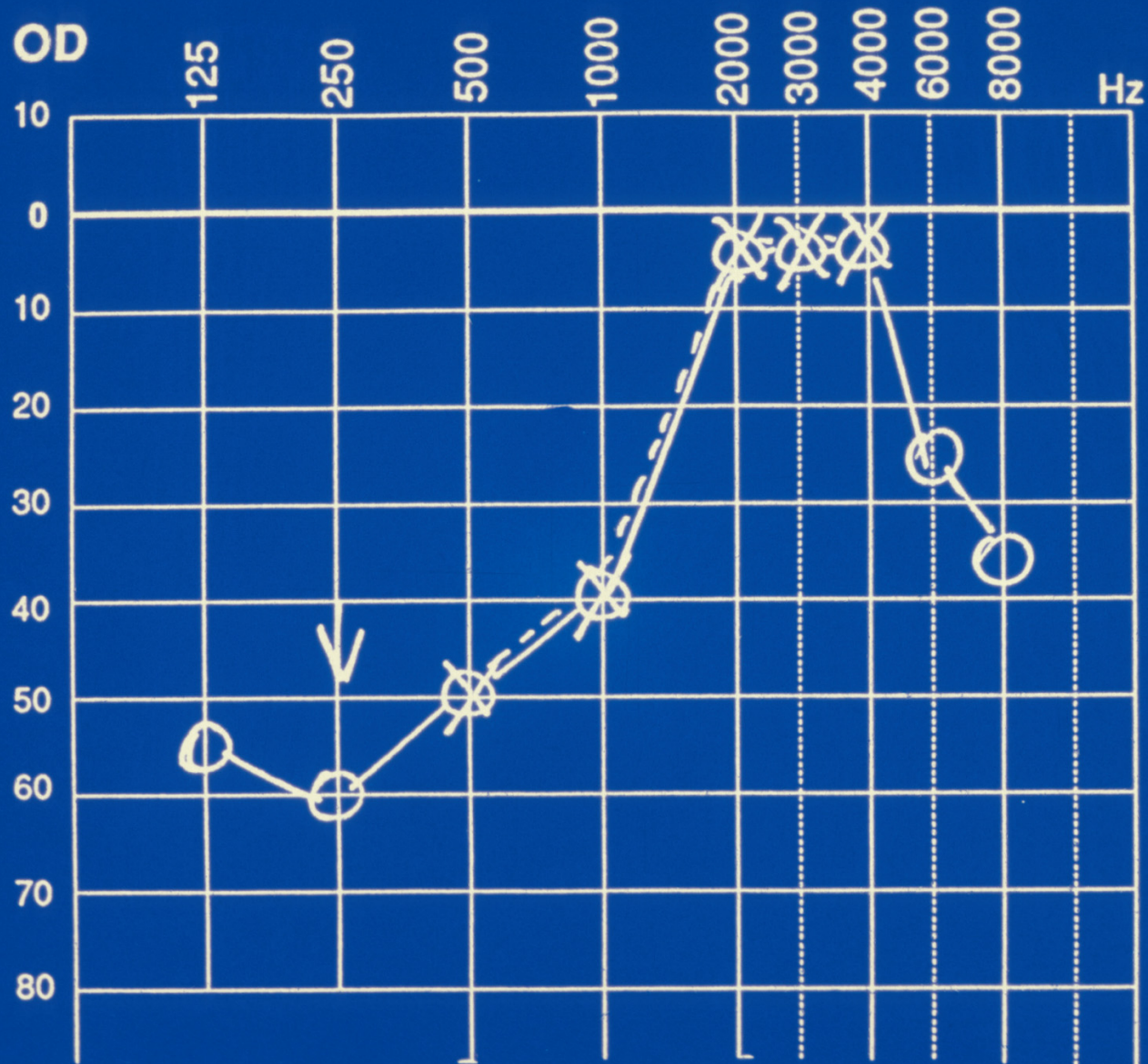
$\alpha\Delta$ 1/2

périphérique
encéphalopathie

occlusion et/ou fuite++



ORL: presbyacousie inversée



traitement

tmt tips

éviter les estrogènes

1ère intention

corticothérapie

- à forte dose SMD 0.5-1 G J1J2J3 (inaugurale/poussée)
- minimiser la dose de prednisone en relais (poussée) $\frac{1}{2}$ mg/kg
- durée totale 12 mois

antiagrégation durée indéfinie

tmt (2)

2^{ème} intention (gravité, poussée récurrente)

IS cyc/aza/mmf

IVIg $\geq$ 3 cures 2 g/kg / 3-4 semaines

3^{ème} intention

rtx 1 g x2

tmt (3)

prothèses auditives
implants cochléaires

suivi

IRM cerveau

audiogramme

CV > AV, FO+/-angio R

les preuves

base épidémiologique

femme jeune

SEX RATIO F / H : 4 / 1

AGE : 30 (2 - 69) ANS

base bioclinique

vasculopathie

+

LCR inflammatoire

base évolutive

- par poussées
- grossesse, post partum
- amélioration clinique in fine
- réversibilité IRM (N lésions diminue au cours du temps)

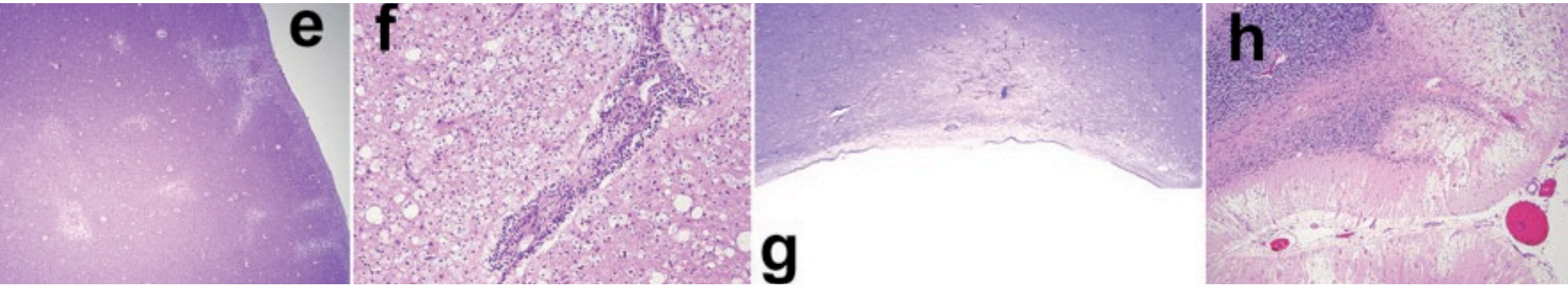
« preuve » histologique 2019

autopsie

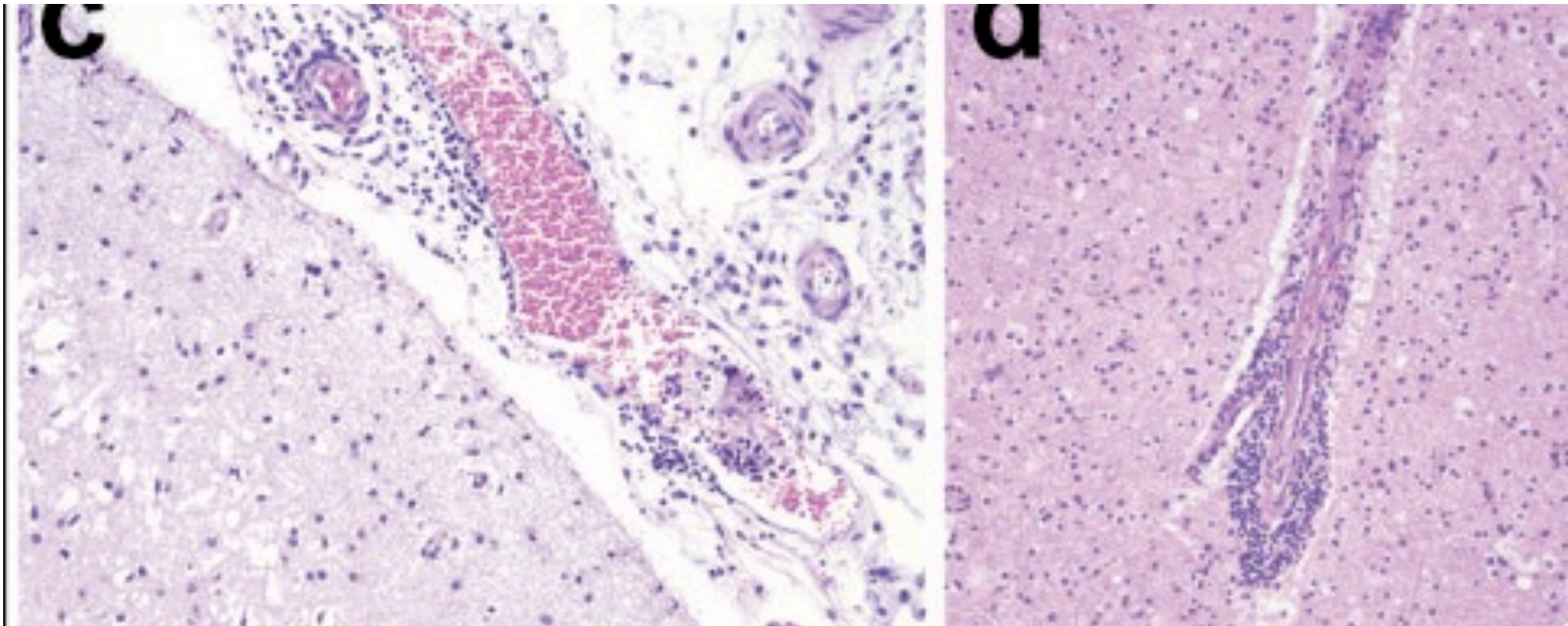
- cerveau quantité ++
- immunophénotypage, ME

bémol: sévérité (atypie++ Susac mortel)

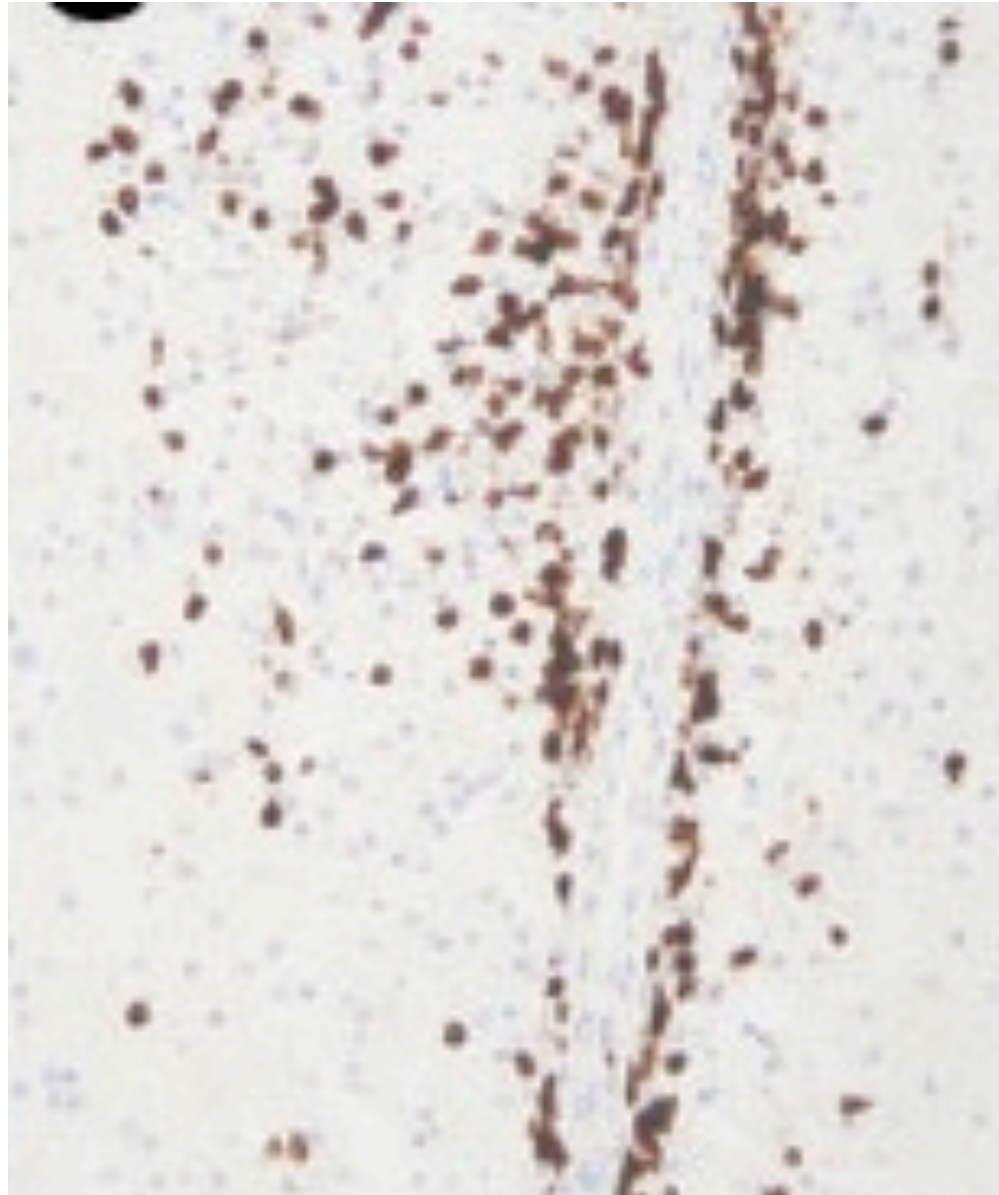
micro-infarctus cortex peri-V, CC, cervelet



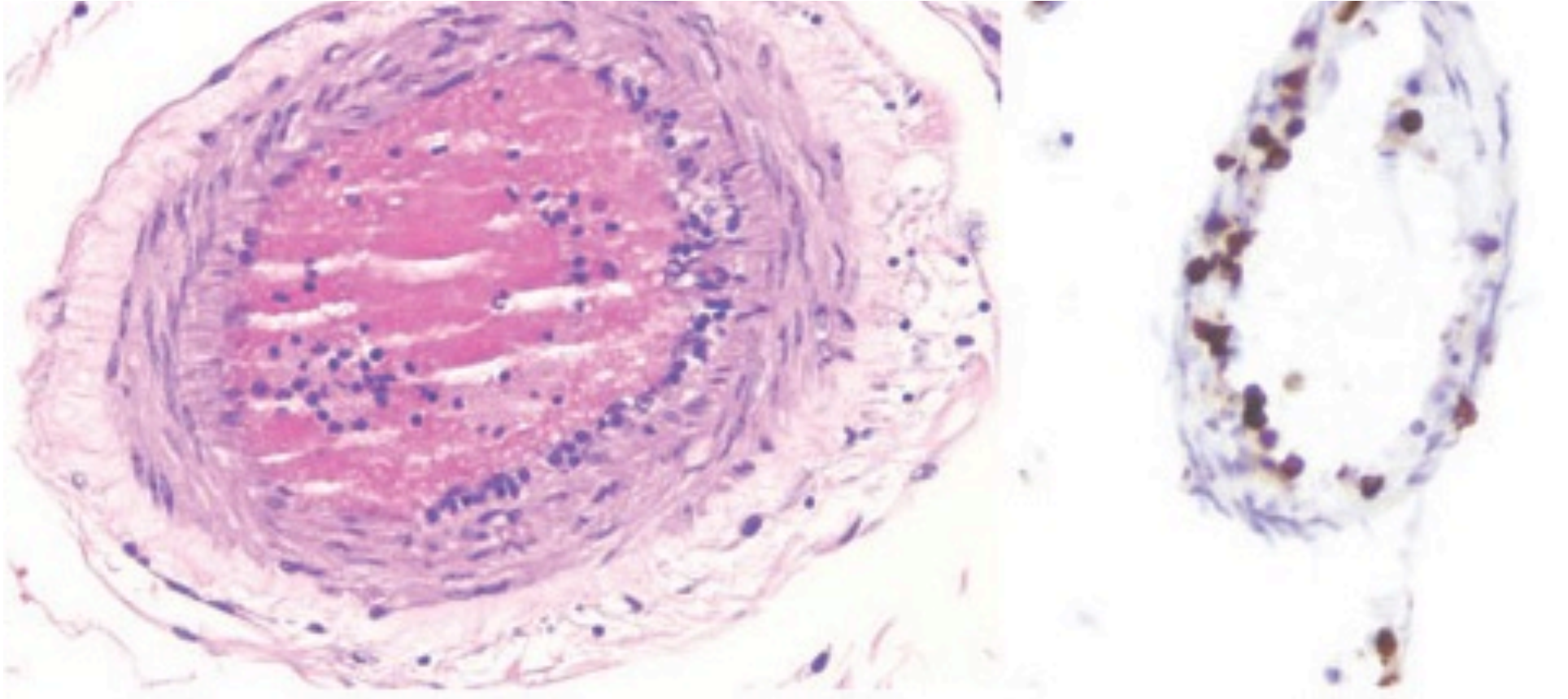
cellules mononucléées
leptoméninge, parenchyme, péri-V



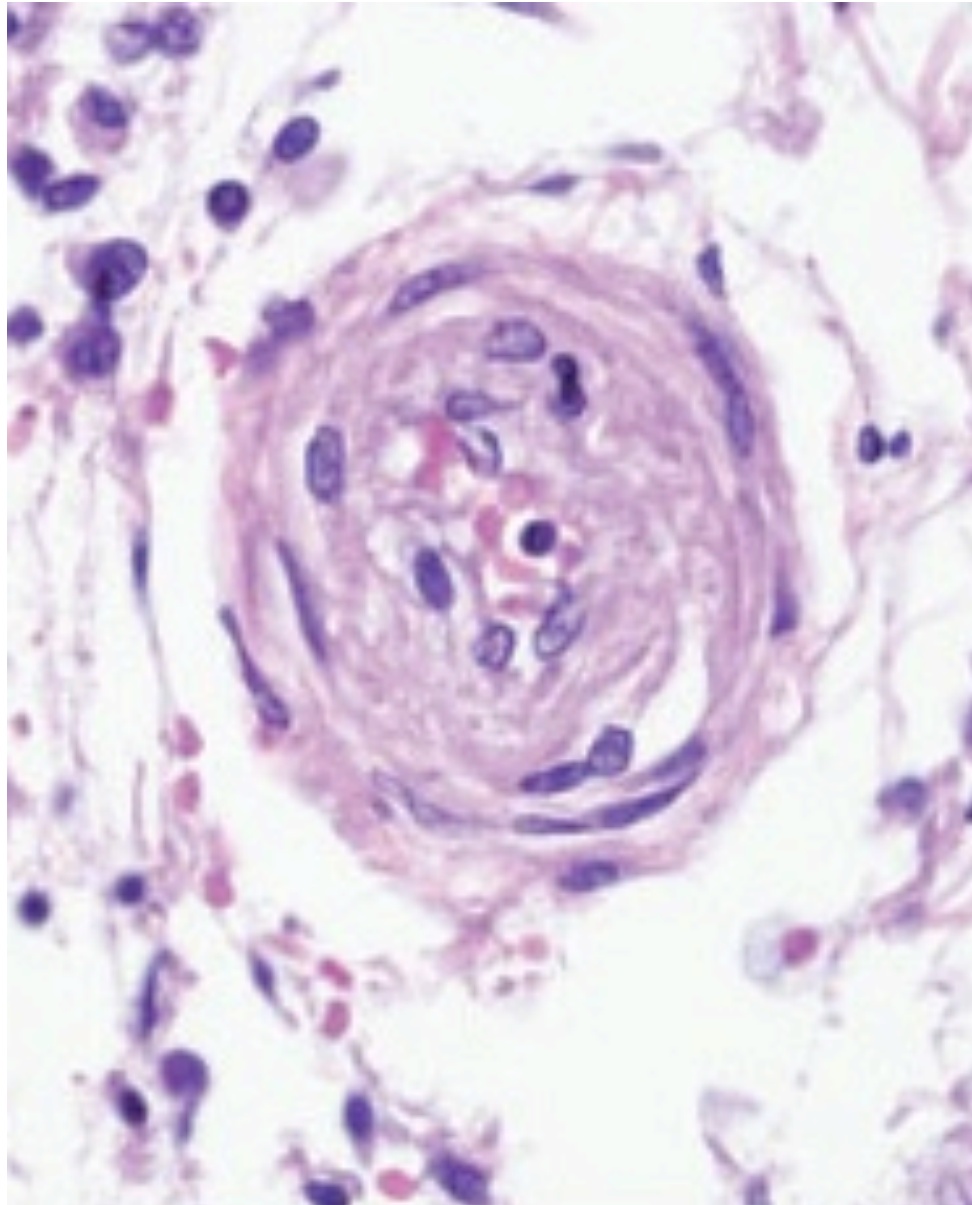
lymphocytes CD3 CD8



cellules mononucléées CD3+ paroi V



artère leptoméningée, fibrose



synthèse

inflammation paroi et peri-V, LT CD8 et M >> LB

non Ac-dep

calibre varié, art leptoméningée - capillaire

thrombi cruoriques, rares

microinfarctus diffus cortex ≠ IRM SB

fuite réversible

vascularite > épithéliopathie primitive

evidence-based, my foot

Parachute use to prevent death and major trauma related to gravitational challenge: systematic review of randomised controlled trials

Gordon C S Smith, Jill P Pell

Abstract

Objectives To determine whether parachutes are effective in preventing major trauma related to gravitational challenge.

Design Systematic review of randomised controlled trials.

Data sources: Medline, Web of Science, Embase, and the Cochrane Library databases; appropriate internet sites and citation lists.

Study selection: Studies showing the effects of using a parachute during free fall.

Main outcome measure Death or major trauma, defined as an injury severity score > 15 .

Results We were unable to identify any randomised controlled trials of parachute intervention.

Conclusions As with many interventions intended to prevent ill health, the effectiveness of parachutes has not been subjected to rigorous evaluation by using randomised controlled trials. Advocates of evidence based medicine have criticised the adoption of interventions evaluated by using only observational data. We think that everyone might benefit if the most radical protagonists of evidence based medicine organised and participated in a double blind, randomised, placebo controlled, crossover trial of the parachute.

accepted intervention was a fabric device, secured by strings to a harness worn by the participant and released (either automatically or manually) during free fall with the purpose of limiting the rate of descent. We excluded studies that had no control group.

Definition of outcomes

The major outcomes studied were death or major trauma, defined as an injury severity score greater than 15.⁶

Meta-analysis

Our statistical approach was to assess outcomes in parachute and control groups by odds ratios and quantified the precision of estimates by 95% confidence intervals. We chose the Mantel-Haenszel test to assess heterogeneity, and sensitivity and subgroup analyses and fixed effects weighted regression techniques to explore causes of heterogeneity. We selected a funnel plot to assess publication bias visually and Egger's and Begg's tests to test it quantitatively. Stata software, version 7.0, was the tool for all statistical analyses.

Results

Our search strategy did not find any randomised controlled trials of the parachute.