



**BRAINWORKS**

The International Inflammatory  
Brain Disease Outcome Study

# **Childhood Primary Central Nervous System Vasculitis Treatment Protocols**

Last updated July 2022

# BrainWorks CNS Vasculitis Protocol

**Non-progressive** large vessel primary CNS vasculitis\*  
 Adjunctive immunosuppression for 3 months

**IV Methyl-prednisolone**

30 mg/kg (max 1g)  
 x \_\_\_ days (3 to 5)

Start date DD/MM/YY

**Prednisolone PO daily**

Dose \_\_\_mg

2 mg/kg (max 60mg)  
 x 3 month tapering monthly  
 (e.g. 60-50-40)

Start date DD/MM/YY

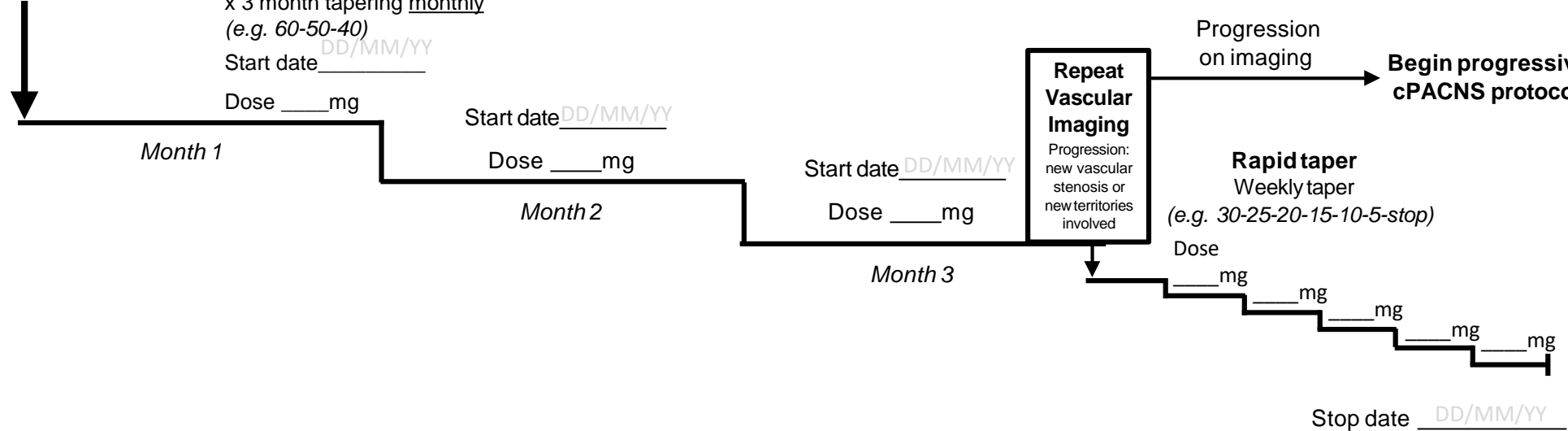
Dose \_\_\_mg

Start date DD/MM/YY

Dose \_\_\_mg

Start date DD/MM/YY

Dose \_\_\_mg



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**Calcium** (1000 mg/day PO)

**Vitamin D** (1000 IU/day PO)

Optional:

**Acyclovir** (80 mg/kg/day PO divided QID, consider renal adjustment)

x \_\_\_\_\_ days (5 to 14, if indicated)

Dose \_\_\_\_\_mg



\* Includes entities of: Angiography-positive, non-progressive primary CNS vasculitis (NPcPACNS), Transient cerebral arteriopathy (TCA), Focal cerebral arteriopathy (FCA), and Post-varicella angiopathy (PVA)

# BrainWorks CNS Vasculitis Protocol

**Progressive** primary large vessel CNS vasculitis\*

Phase 1: Induction Therapy (0-6 months)

### IV Methylprednisolone pulses

30 mg/kg (max 1g)

x \_\_\_\_\_ days (3 to 7)

Start date DD/MM/YY

Dose \_\_\_\_\_ mg

### Prednisone PO daily

2 mg/kg (max 60 mg) tapering monthly

(e.g. 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop)

Start date DD/MM/YY

Dose \_\_\_\_\_ mg

Start date DD/MM/YY

Dose \_\_\_\_\_ mg

Start date DD/MM/YY

Dose \_\_\_\_\_ mg

Start date DD/MM/YY

Dose \_\_\_\_\_ mg

Start date DD/MM/YY

Dose \_\_\_\_\_ mg

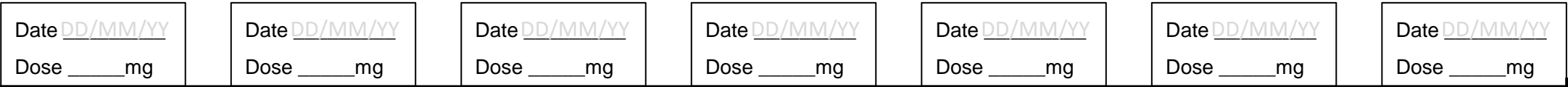
Start date DD/MM/YY

Dose \_\_\_\_\_ mg

Start date DD/MM/YY

Dose \_\_\_\_\_ mg

### Cyclophosphamide (500-750 mg/m<sup>2</sup> monthly x 7 doses)



### Trimethoprim/sulfamethoxazole (PJP prophylaxis dosing)



### Calcium (1000 mg/day PO)



### Vitamin D (1000 IU/day PO)



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\*Angiography-positive, progressive primary CNS vasculitis (PcPACNS)

# BrainWorks CNS Vasculitis Protocol

Progressive primary large vessel CNS vasculitis\*

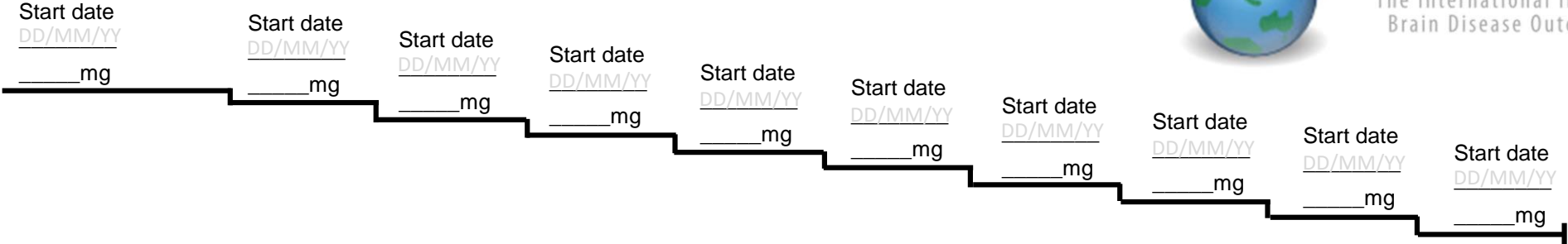
Phase 2: Maintenance Therapy (7-18 months)



### Prednisone PO daily

2 mg/kg (max 60 mg) tapering monthly

(e.g. 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop)



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Choose:

**Mycophenolate mofetil** (800-1200 mg/m<sup>2</sup> divided BID) Dose: \_\_\_\_\_mg Stop date DD/MM/YY

or

**Mycophenolic acid** (500-800 mg/m<sup>2</sup> divided BID) Dose: \_\_\_\_\_mg Stop date DD/MM/YY

or

**Azathioprine** (2-3 mg/kg, max 150 mg, PO daily) Dose: \_\_\_\_\_mg Stop date DD/MM/YY

**Calcium** (1000 mg/day PO)

**Vitamin D** (1000 IU/day PO)

\*Angiography-positive, progressive primary CNS vasculitis (PcPACNS)

# BrainWorks CNS Vasculitis Protocol

Primary **small vessel** CNS vasculitis\*  
Phase 1: Induction Therapy (0-6 months)



### IV Methylprednisolone pulses

30 mg/kg (max 1g)

x \_\_\_\_\_ days (3 to 7)

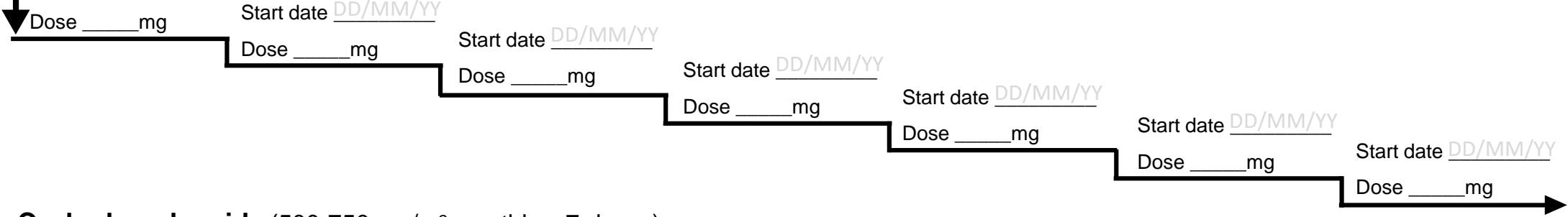
Dose \_\_\_\_\_ mg

Start date DD/MM/YY

### Prednisone PO daily

2 mg/kg (max 60 mg) tapering monthly  
(e.g. 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop)

Start date DD/MM/YY



### Cyclophosphamide (500-750 mg/m<sup>2</sup> monthly x 7 doses)

Date <u>DD/MM/YY</u> Dose _____ mg	Date <u>DD/MM/YY</u> Dose _____ mg	Date <u>DD/MM/YY</u> Dose _____ mg	Date <u>DD/MM/YY</u> Dose _____ mg	Date <u>DD/MM/YY</u> Dose _____ mg	Date <u>DD/MM/YY</u> Dose _____ mg	Date <u>DD/MM/YY</u> Dose _____ mg
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### Trimethoprim/sulfamethoxazole (prophylaxis dosing)



### Calcium (1000 mg/day PO)



### Vitamin D (1000 IU/day PO)



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\*Angiography-positive, progressive primary CNS vasculitis (PcPACNS)

# BrainWorks CNS Vasculitis Protocol

Primary **small vessel** CNS vasculitis\*

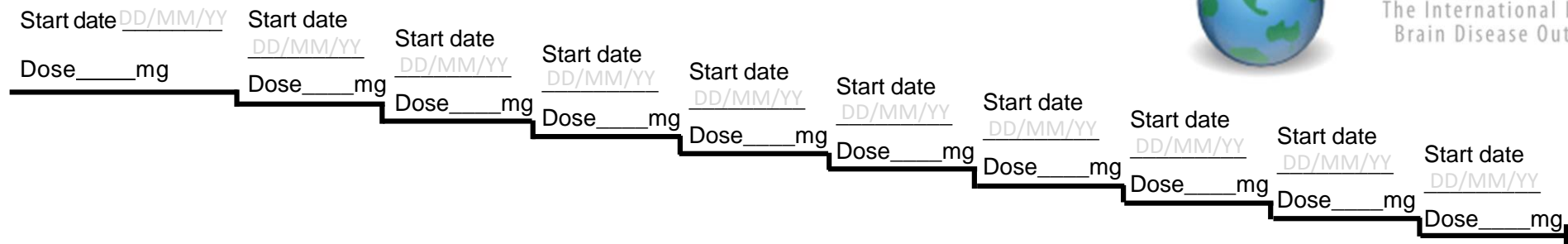
Phase 2: Maintenance Therapy (6-18 months)



**Prednisone PO daily**

Tapering monthly\*\*

(e.g. 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop)



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Choose:

Stop date DD/MM/YY

**Mycophenolate mofetil** (800-1200 mg/m<sup>2</sup> divided BID) Dose: \_\_\_\_\_

Stop date DD/MM/YY

or

**Mycophenolic acid** (500-800 mg/m<sup>2</sup> divided BID) Dose: \_\_\_\_\_

Stop date DD/MM/YY

or

**Azathioprine** (2-3 mg/kg, max 150 mg, PO daily) Dose: \_\_\_\_\_

Stop date DD/MM/YY

**Calcium** (1000 mg/day PO)

**Vitamin D** (1000 IU/day PO)

\*Angiography-positive, progressive primary CNS vasculitis (PcPACNS)

# BrainWorks CNS Vasculitis Protocol

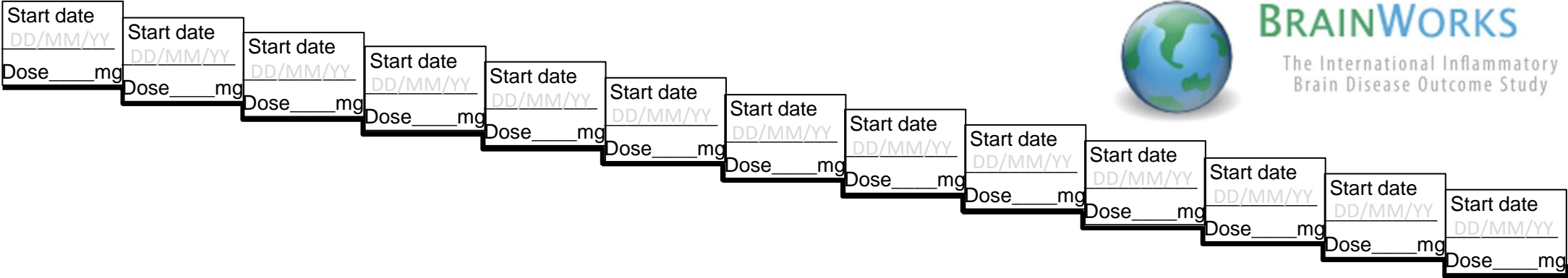
Refractory primary CNS vasculitis\*

Phase 1: Induction 0 - 6 months

**Prednisone PO daily**

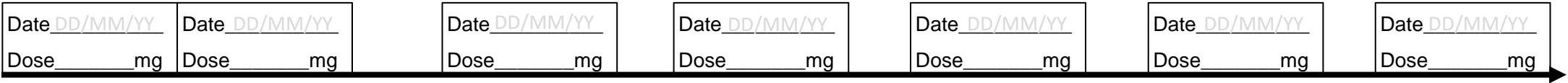
2 mg/kg (max 60 mg) tapering every 2 weeks

(e.g. 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop)

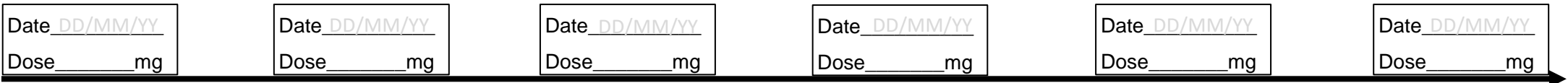


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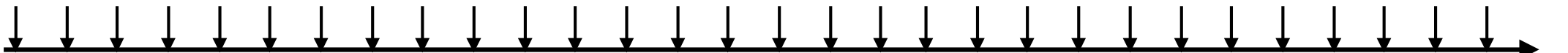
**Infliximab** (7.5 mg/kg IV, at diagnosis and after 2 weeks, then monthly following)



**IVIg** (500-750 mg/m<sup>2</sup> monthly)



**Methotrexate** (10-15 mg PO/subcutaneously weekly) Dose: \_\_\_\_\_ mg



**Calcium** (1000 mg/day PO)



**Vitamin D** (1000 IU/day PO)



**Folic Acid** (1 mg/day PO)



\* Failure to respond to induction therapy

# BrainWorks CNS Vasculitis Protocol

Refractory primary CNS vasculitis\*  
Phase 2: Maintenance (7 - 12 months)



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## Infliximab (7.5 mg/kg IV monthly)

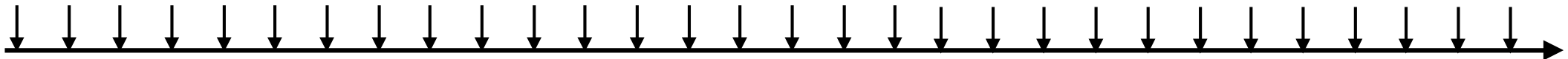
Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg
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Optional:

## IVIG (500-750 mg/m<sup>2</sup> monthly)

Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg	Date <u>DD/MM/YY</u> Dose _____mg
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## Methotrexate (10-15 mg PO/subcutaneously weekly) Dose: \_\_\_\_\_mg



## Calcium (1000 mg/day PO)



## Vitamin D (1000 IU/day PO)



## Folic Acid (1 mg/day PO)



\* Failure to respond to induction therapy



# BrainWorks CNS Vasculitis Protocol

Disease activity **flare** of primary CNS vasculitis\*

Treat for 6 months

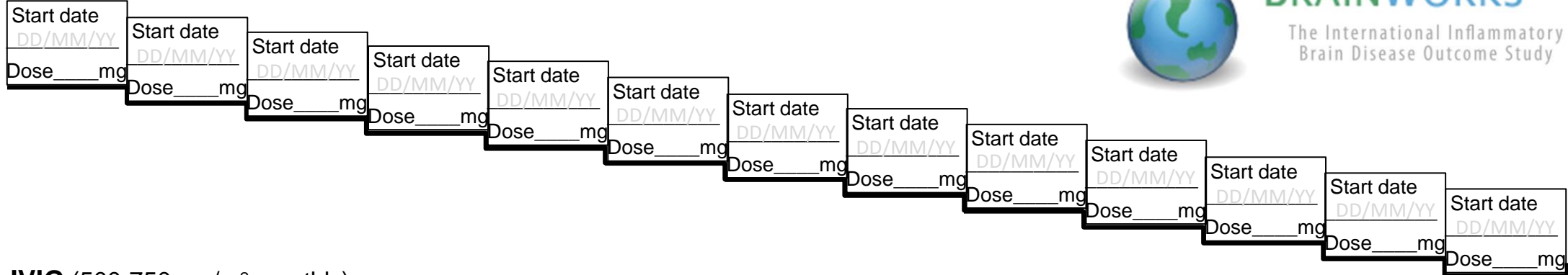


## Minor flare

### Prednisone PO daily

2 mg/kg (max 60 mg) tapering every 2 weeks

(e.g. 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop)



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## IVIG (500-750 mg/m<sup>2</sup> monthly)



**Mycophenolate mofetil** (800-1200 mg/m<sup>2</sup> divided BID) Dose: \_\_\_\_\_ mg Stop date DD/MM/YY

or

**Mycophenolic acid** (500-800 mg/m<sup>2</sup> divided BID) Dose: \_\_\_\_\_ mg Stop date DD/MM/YY

**Calcium** (1000 mg/day PO)

**Vitamin D** (1000 IU/day PO)

## Major flare

Choose as per clinical presentation:

Repeat Phase 1: Induction Therapy protocol

or

Proceed to Refractory Primary CNS Vasculitis Therapy protocol

\* Defined as recurrence of symptoms relative to baseline functional status