



**BRAINWORKS**  
The International Inflammatory  
Brain Disease Outcome Study

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

Last updated December 2014

# BrainWorks CNS Vasculitis Protocol

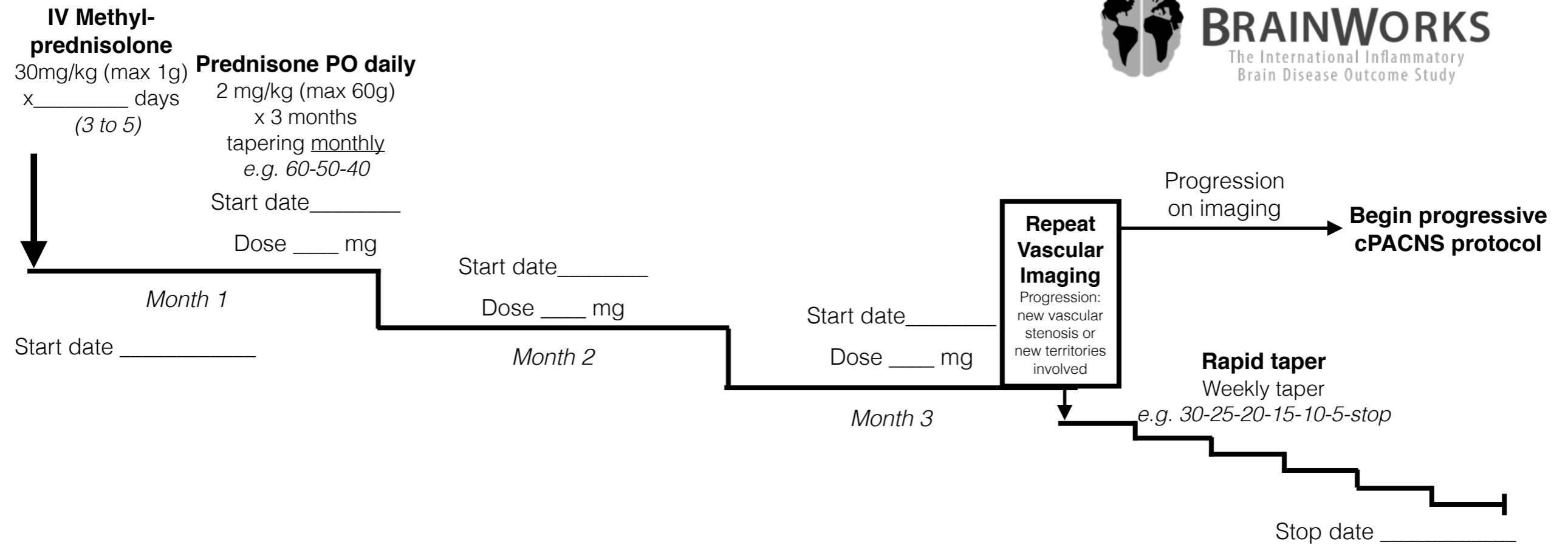
**Non-progressive** large vessel primary CNS vasculitis\*

Adjunctive immunosuppression for 3 months



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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)



\* 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

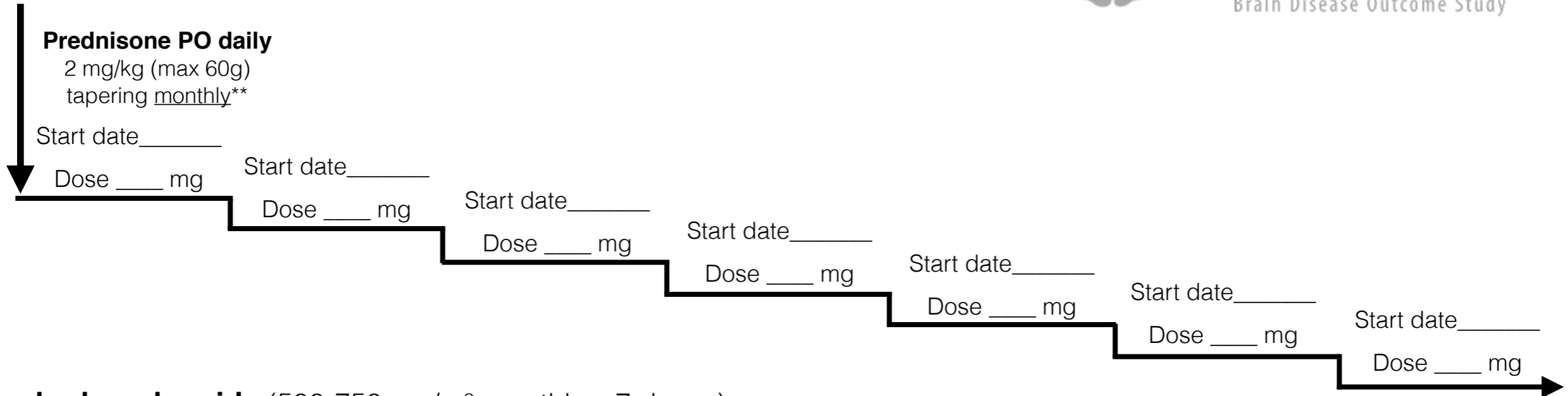
30mg/kg (max 1g)  
x \_\_\_\_\_ days (3-7)

Start date \_\_\_\_\_

## Prednisone PO daily

2 mg/kg (max 60g)  
tapering monthly\*\*

Start date \_\_\_\_\_



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

Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____
Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg

## 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)

\*\* Example prednisone taper schedule: 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop

# BrainWorks CNS Vasculitis Protocol

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

Phase 2: Maintenance Therapy (7-18 months)



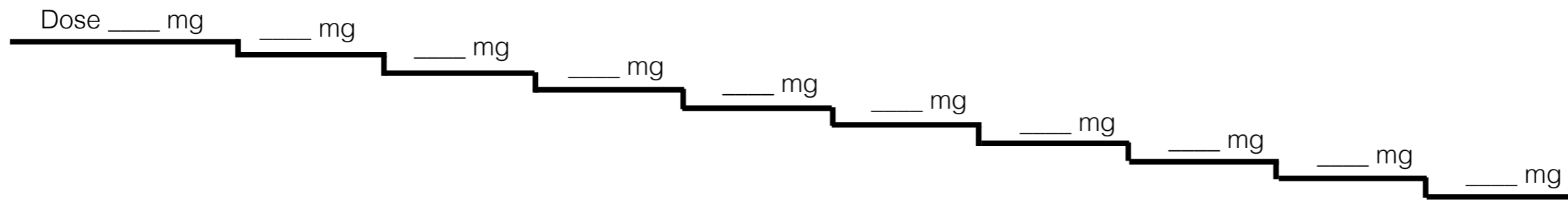
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### Prednisone PO daily

Tapering monthly\*\*

Start date \_\_\_\_\_



Choose:

End of taper date \_\_\_\_\_

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

or

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

or

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

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

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

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

\*\* Example prednisone taper schedule: 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop

# BrainWorks CNS Vasculitis Protocol

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



## IV Methylprednisolone pulses

30mg/kg (max 1g)  
x \_\_\_\_\_ days (3-7)

Start date \_\_\_\_\_



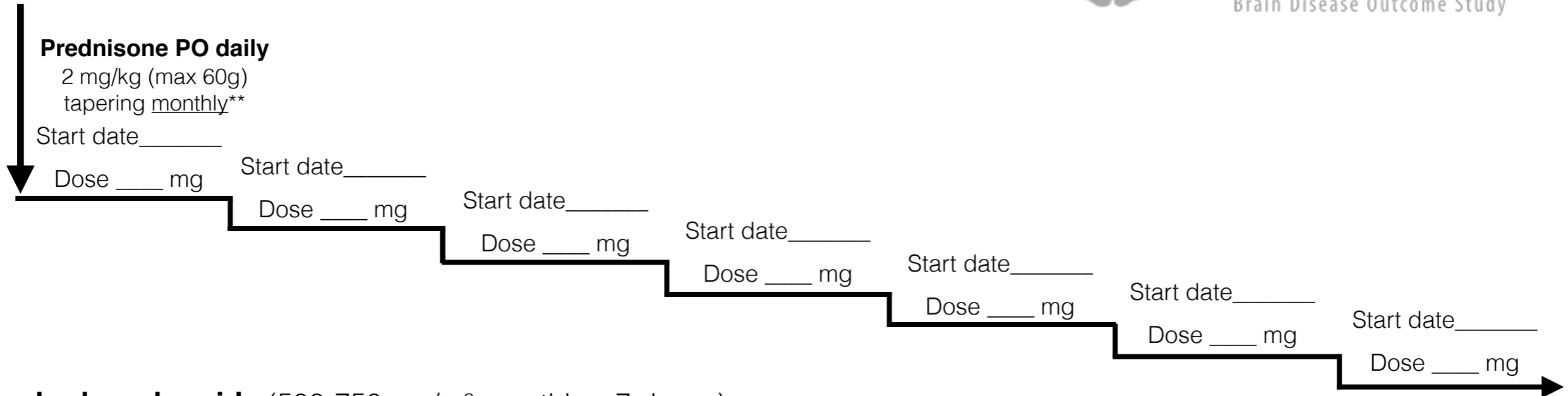
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## Prednisone PO daily

2 mg/kg (max 60g)  
tapering monthly\*\*

Start date \_\_\_\_\_



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

Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____
Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg

## Trimethoprim/sulfamethoxazole (prophylaxis dosing)



## Calcium (1000 mg/day PO)



## Vitamin D (1000 IU/day PO)



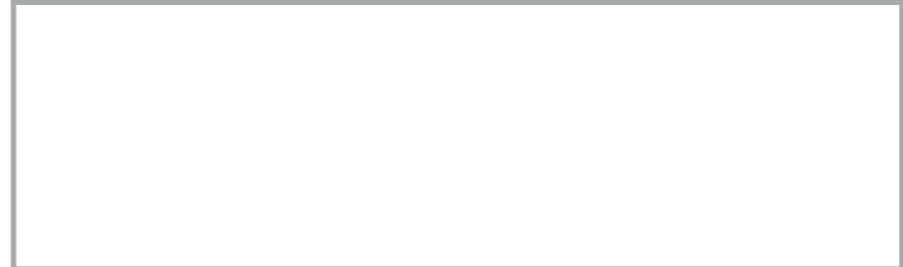
\*Angiography-negative, brain-biopsy positive small vessel primary CNS vasculitis (SVcPACNS)

\*\* Example prednisone taper schedule: 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop

# BrainWorks CNS Vasculitis Protocol

Primary **small vessel** CNS vasculitis\*

Phase 2: Maintenance Therapy (6-18 months)



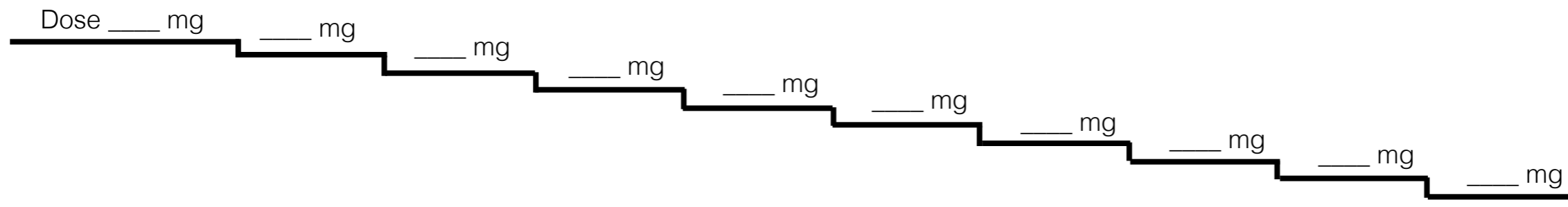
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**Prednisone PO daily**

Tapering monthly\*\*

Start date \_\_\_\_\_



Choose:

Stop date \_\_\_\_\_

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

Stop date \_\_\_\_\_

*or*

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

Stop date \_\_\_\_\_

*or*

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

Stop date \_\_\_\_\_

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

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

\*Angiography-negative, small vessel primary CNS vasculitis (SVcPACNS)

\*\* Example prednisone taper schedule: 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop

# BrainWorks CNS Vasculitis Protocol

**Refractory** primary CNS vasculitis\*

Phase 1: Induction 0 - 6 months

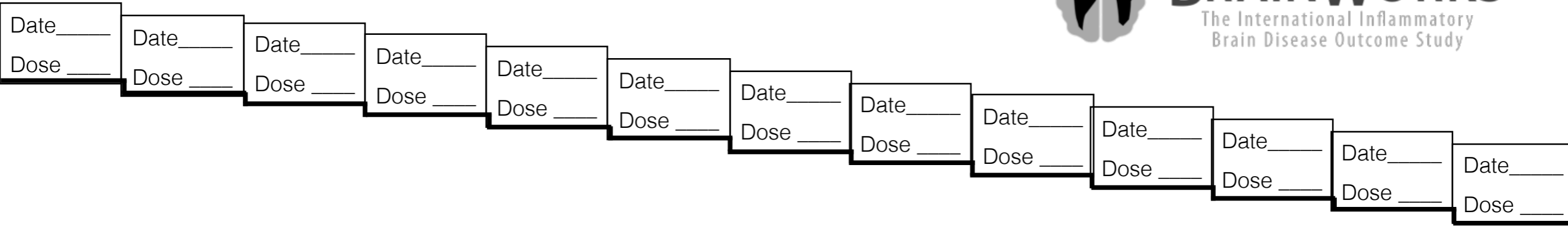


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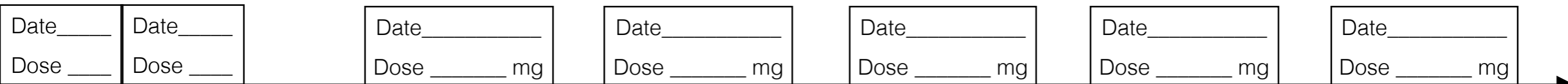
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**Prednisone PO daily**

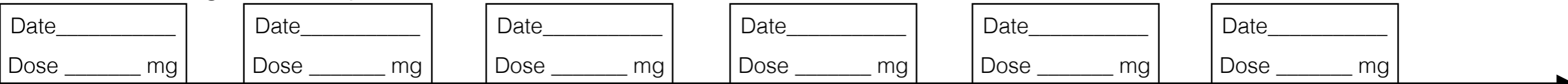
2 mg/kg (max 60g)  
tapering biweekly\*\*



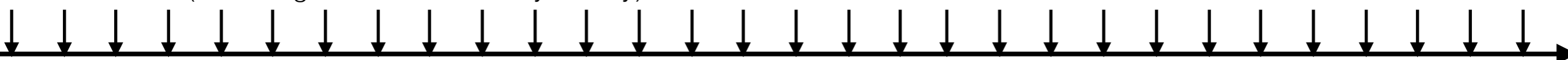
**Infliximab** (5-7.5 mg/kg IV, at diagnosis and after 2 weeks, then monthly)



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



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



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



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



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



\* Failure to respond to induction therapy

\*\* Example prednisone taper schedule: 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop

# BrainWorks CNS Vasculitis Protocol

**Refractory** primary CNS vasculitis\*

Phase 2: Maintenance (7 - 12 months)



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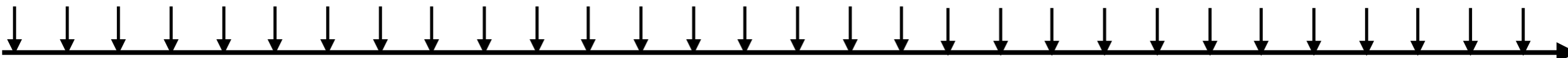
**Infliximab** (5-7.5 mg/kg IV monthly)

Date _____	Date _____	Date _____	Date _____	Date _____	Date _____
Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg

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

Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____
Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg

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



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



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



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



\* Failure to respond to induction therapy

\*\* Example prednisone taper schedule: 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop



# BrainWorks CNS Vasculitis Protocol

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

Treat for 6 months



## Minor flare

### Prednisone PO daily

2 mg/kg (max 60g)  
tapering biweekly\*\*



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Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____	Date _____
Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____	Dose _____

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

Date _____	Date _____	Date _____	Date _____	Date _____	Date _____
Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg	Dose _____ mg

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

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

**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

\*\* Example prednisone taper schedule: 60 mg/day, 50, 40, 30, 25, 20, 17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop