



BRAINWORKS
The International Inflammatory
Brain Disease Outcome Study

Childhood Primary Central Nervous System Vasculitis Treatment Protocols

Last updated December 2014

BrainWorks CNS Vasculitis Protocol

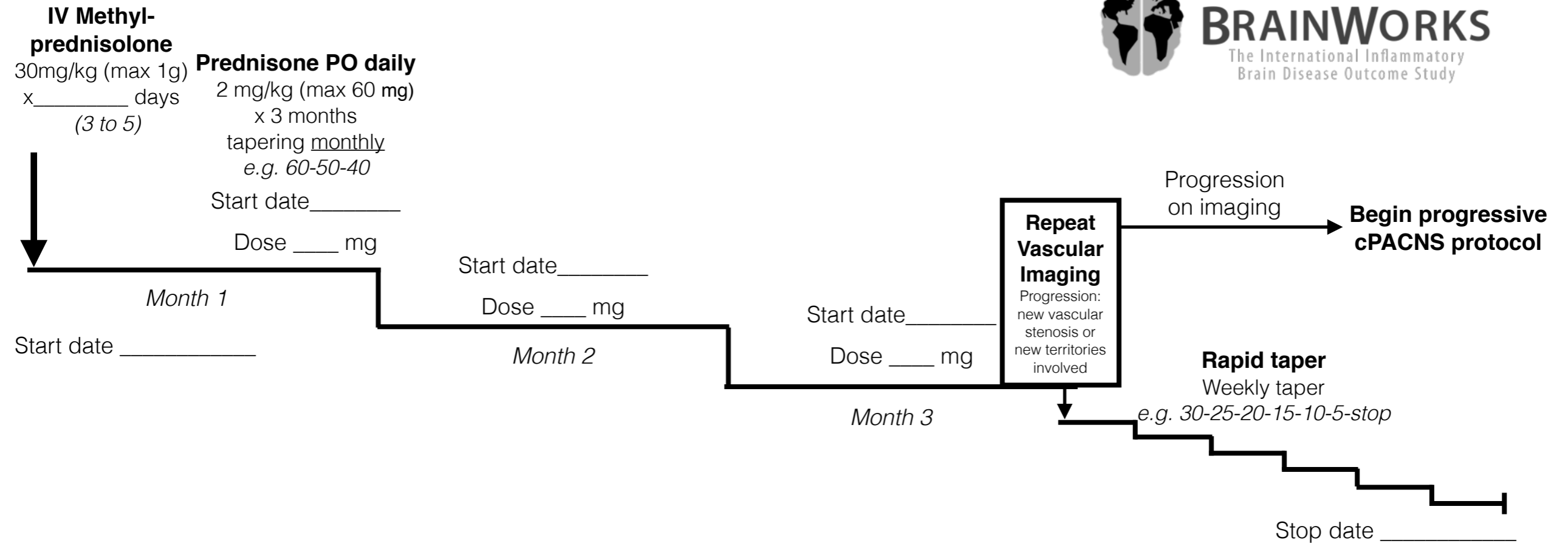
Non-progressive large vessel primary CNS vasculitis*

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

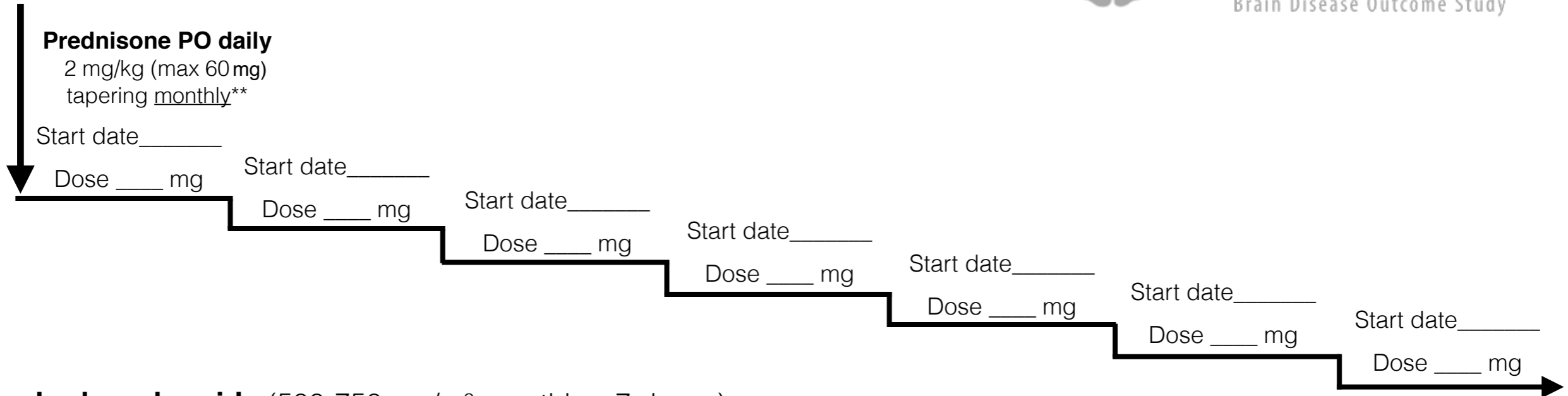
30mg/kg (max 1g)
x _____ days (3-7)

Start date _____

Prednisone PO daily

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

Start date _____



Cyclophosphamide (500-750 mg/m² 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)



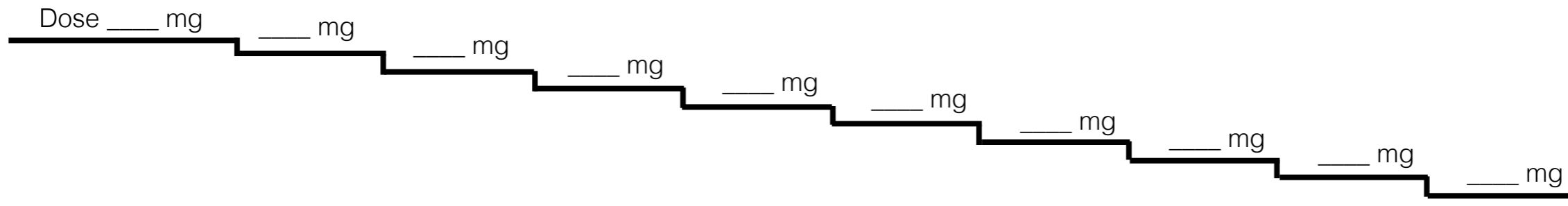
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Prednisone PO daily

Tapering monthly**

Start date _____



Choose:

Mycophenolate mofetil (800-1200 mg/m² divided BID) Dose: _____ Stop date _____

or

Mycophenolic acid (500-800 mg/m² 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 _____



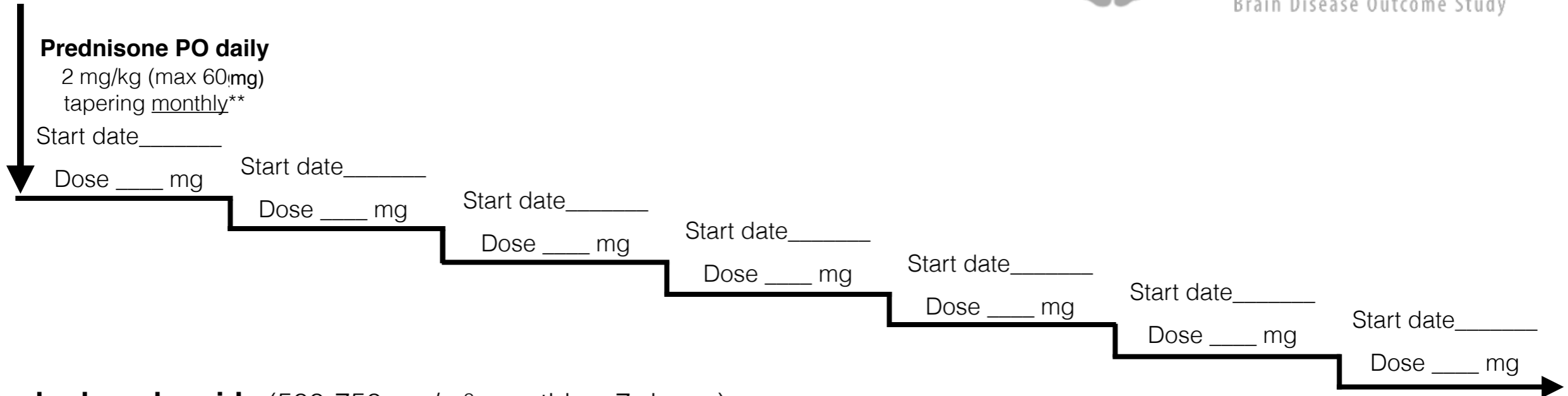
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Prednisone PO daily

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

Start date _____



Cyclophosphamide (500-750 mg/m² 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)



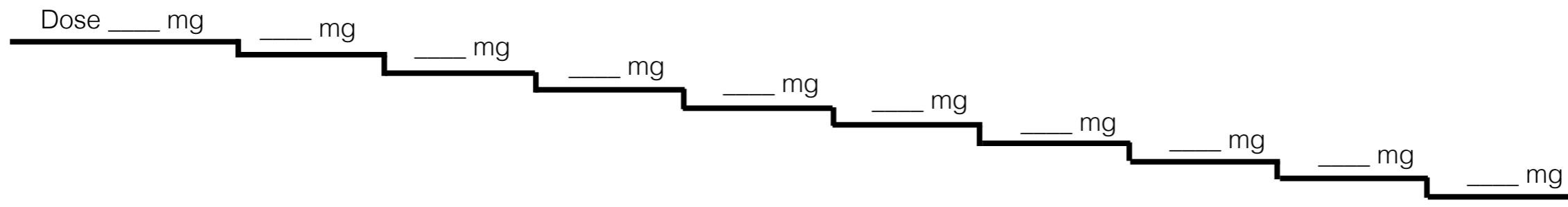
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Prednisone PO daily

Tapering monthly**

Start date _____



Choose:

Stop date _____

Mycophenolate mofetil (800-1200 mg/m² divided BID) Dose: _____

Stop date _____

or

Mycophenolic acid (500-800 mg/m² 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

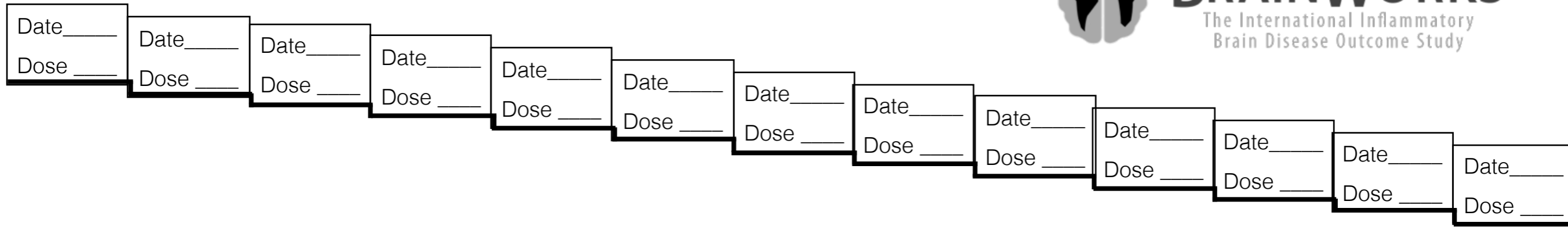


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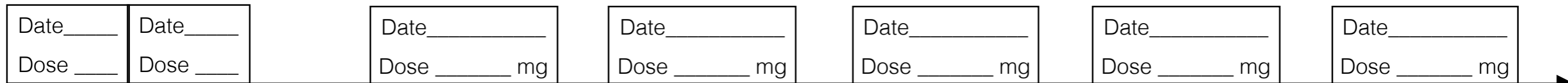
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Prednisone PO daily

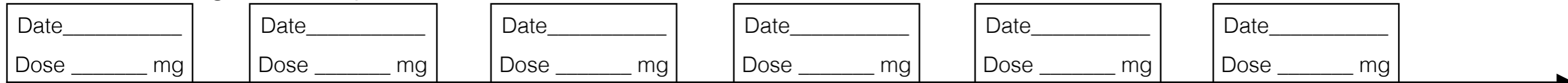
2 mg/kg (max 60 mg)
tapering biweekly**



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



IVIg (500-750 mg/m² 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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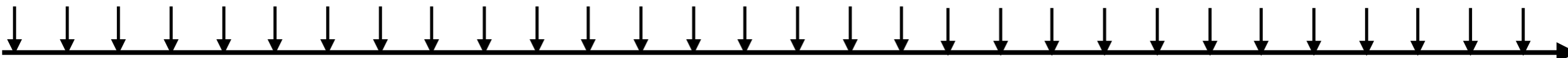
Infliximab (5-7.5 mg/kg IV monthly)

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

Optional: **IVIg** (500-750 mg/m² 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 60 mg)
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/m² monthly)

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

Mycophenolate mofetil (800-1200 mg/m² divided BID) Dose: _____ Stop date _____

or
 Mycophenolic acid (500-800 mg/m² 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