

Sodium Phenylbutyrate Intravenous Administration Guideline—Page 1 of 3

—Prescribing of Loading doses and Continuous Infusions

Loading dose (if required)	250mg/kg over 90 minutes
Maintenance dose (continuous infusion)	250-500mg/kg/day
Step 1 — Calculate daily requirements in grams [i.e. loading dose (if required) + maintenance dose]	e.g. 4kg patient requires 250mg/kg load followed by 250mg/kg/day maintenance (continuous infusion) [(4x250 = 1000mg load) + (4x250 = 1000mg daily maintenance) = 2000mg = 2g]
Step 2 — Read off the preparation of solution guide (page2) to determine the formula for the prescription	Using the above example, 2g Sodium Phenylbutyrate will be prepared, making up to 40mL final volume with Glucose 10%w/v.
Step 3 — Complete the prescription with full details for the patient – i.e. formula, +/- loading dose and maintenance dose. A new prescription is written if the maintenance dose changes.	See completed model prescription below for example

Sodium Phenylbutyrate Intravenous Administration Guideline—Page 2 of 3—Preparation of Solution						
Total calculated daily dose (g)	Dose to be prepared (g)	Volume (mL) of neat Sodium Phenylbutyrate (using 1g in 5mL or 2g in 10mL)	Volume (mL) of Glucose 10%w/v diluent	Final Volume (mL)	Final concentration of Sodium Phenylbutyrate (%w/v = number of g in 100mL)	Preparation Instructions
0-2g	2	10	30	40	5%w/v	Use syringe
>2-4g	4	20	60	80	5%w/v	Use Buretrol

Model Prescription e.g. 4kg patient

INTRAVENOUS INFUSIONS

INTRAVENOUS INFUSIONS - IV fluids and Medication

Remember to check allergy section at the front of the kardex before prescribing
Where an amendment is necessary, please rewrite the prescription IN FULL.

Affix Patient Identifier

Infusion Fluid or Base Solution	GLUCOSE 10% 4/10	Final Volume (mL)	40mL	Date Prescribed	10/2/20	Prep. Check	Date	Time	Pump Check	Date	Time
Medication or Electrolyte to be added	Name: SODIUM PHENYLBUTYRATE Quantity: 2g			12345 Reg. No.	Joe Bloggs Sleep No. 110	① JC MK	3/2/20 0900		JC MK	3/2/20 0910	
Rate	① Loading Dose 1g (= 250mg/kg) over 90 mins.			2		②			JC MK	3/2/20 1040	
Additional Instructions	then ② maintenance dose 250mg/kg/day		Batch no./EXP (if applicable)	4							
			Date Cancelled	<div style="background-color: #e91e63; color: white; padding: 5px; display: inline-block;"> CANCELLED (complete with full prescriber details) </div>							

Sodium Phenylbutyrate Intravenous Administration Guideline

—Page 2 of 3—Preparation of Solution

Total calculated daily dose (g)	Dose to be prepared (g)	Volume (mL) of neat Sodium Phenylbutyrate (using 1g in 5mL or 2g in 10mL)	Volume (mL) of Glucose 10%w/v diluent	Final Volume (mL)	Final concentration of Sodium Phenylbutyrate (%w/v = number of g in 100mL)	Preparation Instructions
0-2g	2	10	30	40	5%w/v	Use syringe
>2-4g	4	20	60	80	5%w/v	Use Buretrol
>4-6g	6	30	90	120	5%w/v	Use Buretrol
>6-8g	8	40	120	160	5%w/v	Use Sterile 250ml empty bags* ¹
>8-10g	10	50	150	200	5%w/v	Use Sterile 250ml empty bags* ¹
>10-12g	12	60	180	240	5%w/v	Use Sterile 250ml empty bags* ¹
>12-14g	14	70	210	280	5%w/v	Withdraw 320mL from a 500mL* ² bag and replace with 70mL of Sodium Phenylbutyrate concentrate (1g in 5mL or 2g in 10mL) {530 - 320 + 70 = 280}
>14-16g	16	80	240	320	5%w/v	Withdraw 290mL from a 500mL* ² bag and replace with 80mL of Sodium Phenylbutyrate concentrate (1g in 5mL or 2g in 10mL) {530 - 290 + 80 = 320}
>16-18g	18	90	270	360	5%w/v	Withdraw 260mL from a 500mL* ² bag and replace with 90mL of Sodium Phenylbutyrate concentrate (1g in 5mL or 2g in 10mL) {530 - 260 + 90 = 360}
>18-20g	20	100	300	400	5%w/v	Withdraw 230mL from a 500mL* ² bag and replace with 100mL of Sodium Phenylbutyrate concentrate (1g in 5mL or 2g in 10mL) {530 - 230 + 100 = 400}
>20-22g	22	110	330	440	5%w/v	Withdraw 200mL from a 500mL* ² bag and replace with 110mL of Sodium Phenylbutyrate concentrate (1g in 5mL or 2g in 10mL) {530 - 200 + 110 = 440}
>22-24g	24	120	360	480	5%w/v	Withdraw 170mL from a 500mL* ² bag and replace with 120mL of Sodium Phenylbutyrate concentrate (1g in 5mL or 2g in 10mL) {530 - 170 + 120 = 480}
>24g* ³	26	130	390	520	5%w/v	Withdraw 140mL from a 500mL* ² bag and replace with 130mL of Sodium Phenylbutyrate concentrate (1g in 5mL or 2g in 10mL) {530 - 140 + 130 = 520}

*¹ See Instruction Sheet for Drug Infusion Preparation via Empty Sterile 250ml Bag

*² 500mL bags are determined to have an average volume of 530mL

*³ For all patients requiring >24g per day, make up a bag of 26g in 520ml final volume as directed.

If the total daily dose is >26g, this will have to be prepared a second time within the 24 hour period.

Sodium Phenylbutyrate Intravenous Administration Guideline—Page 3 of 3

—Rate calculation and Programming the Infusion Pump

Step 1 —Prepare the infusion as per prescription and preparation of solution guide (page 2)	e.g. previous example of 4kg patient requiring 2g (including loading dose) - prepare 2g in 40mL final volume Glucose 10%w/v.
Step 2 —Choose appropriate pump (Infusomat® or Perfusor®)	e.g. using previous example volume is ≤50mL so the appropriate pump is Perfusor®
Step 3 —If loading dose is required, calculate the required volume to be infused (<i>if no loading dose is needed proceed to step 5</i>)	As from table below, a 250mg/kg loading dose for a 4kg patient will require 20mL [VTBI = 5 x Wt]
Step 4 —Programme the pump to administer “Sodium Phenylbutyrate Load”. (<i>This is found in the care unit “Metabolic Agents”</i>)	Volume to be infused (VTBI) according to the pump should match your calculation
Step 5 —For the maintenance dose, using the table, calculate the default rate (mL/hour) for the default start dose	(0.417 x Wt)mL/hour = 500mg/kg/day e.g. (0.417 x 4)mL/hour = 500mg/kg/day 1.67mL/hour = 500mg/kg/day
Step 6 —If the required dose is different from the default start dose, calculate the rate (mL/hour) for the required dose using the “rate calc formula”	e.g. Using previous example where required maintenance dose for 4kg patient is 250mg/kg/day = $\frac{250 \times 1.67}{500}$ = 0.84mL/hour
Step 7 —Programme the pump to administer “Sodium Phenylbutyrate Maintenance”.	The flow rate (mL/hour) according to the pump should match your calculation

Metabolic Agents : CONTINUOUS INFUSIONS AND LOADING DOSES STANDARD CONCENTRATION INFUSION DRUG LIBRARY					Rate calc (mL/hour) = $\frac{\text{Required Dose} \times \text{Default Rate (ml/hr)}}{\text{Default Start Dose}}$		
Drug	Category	Weight Band	SCI (Normal)	Diluent	Usual Dose Range	Default Dose and Rate Calculator	
						All Weights in kg - rounding can occur	
						Default Start Dose	Default Rate (mL/hr)
Sodium Phenylbutyrate Load <i>(In own care-unit on pumps - "Metabolic Agents")</i> <i>Scroll past weight-bands and choose "Change care"</i>		All	2g/40mL (Perfusor) or 50mg/mL (Infusomat) <i>see protocol</i>	Glucose 10% w/v	250mg/kg over 90 mins	250mg/kg	VTBI (mL) = 5 x Wt
Sodium Phenylbutyrate Maintenance <i>(In own care-unit on pumps - "Metabolic Agents")</i> <i>Scroll past weight-bands and choose "Change care unit"</i>		All	2g/40mL (Perfusor) or 50mg/mL (Infusomat) <i>see protocol</i>	Glucose 10% w/v	250-500mg/kg/24hours	500mg/kg/24hours	0.417 x Wt