INFANTILE SPASMS: HIGH DOSE ORAL PREDNISOLONE

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- High dose oral prednisolone is used in the treatment of a neurological syndrome characterised by infantile spasms (seizures) and psychomotor retardation, often associated with a characteristic EEG pattern known as hypsarrhythmia. This condition usually occurs in the first year of life, with a peak onset of between 3 – 6 months.
- The spasms may occur in clusters on multiple occasions throughout the day and are usually characterised by sudden muscular contractions in which the head is flexed, the arms extended and the legs drawn upwards or extended. The child may also turn red in the face, become pale or cyanotic. Extension of the limbs and trunk may also be apparent during the spasm. The aim of treatment is to control seizures and arrest any further regression of developmental milestones.
- **Hypertension** is a serious side effect of high dose oral prednisolone requiring strict monitoring of blood pressure.
- Blood pressure for children receiving high dose prednisolone **must** be measured using a manual cuff on the **same** arm each time.
- Relative adrenocortical insufficiency may continue after the high dose oral prednisolone treatment has been ceased.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
Date Effective:	1st December 2013	**Review Period:** 3 years
Team Leader:	CNC neurology Liaison	**Area/Dept:** Neurology & Neurosurgery

Date of Publishing: 17 December 2013 12:36 PM Date of Printing: 17 December 2013

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CHANGE SUMMARY

- The CHW Practice Guideline “Tetracosactrin Depot "Synacthen Depot": Administration and Management of Infantile Spasms” has been rescinded and replaced by this document.
- The review only involved formatting changes and updating references.

READ ACKNOWLEDGEMENT

- All clinical staff involved in the care of children with infantile spasms who are being treated with high dose oral prednisolone are to read and acknowledge they understand the contents of this document.

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Rationale

- To facilitate early detection and management of side effects.
- To facilitate timely referral to the Neuroscience/Epilepsy Clinical Nurse Consultant and maximise the education, support and preparation of the child and family for discharge.
- To facilitate the safe and effective administration of high dose oral prednisolone in children with infantile spasms.
- To facilitate the transition of care for children receiving high dose oral prednisolone from the acute setting to the community setting, with adequate education and support.

General Principles

- Prescribing of the high dose oral steroids is based on the United Kingdom Infantile Spasms Study[2].
- At the commencement of treatment, blood pressure is measured once a day using a manual cuff and sphygmomanometer.
- One of the most important factors in accurately measuring blood pressure is the use of an appropriately sized cuff. Cuff size, which refers to the inner inflatable bladder and not the covering material, must be chosen correctly. When measuring cuff size, select a cuff that is large enough to cover the upper arm down to the cubital fossa.
- Cuff size and the designated arm for blood pressure measurement are documented in the patient's notes.
- A manual sphygmomanometer may be supplied upon discharge if either the appropriate cuff size or a manual sphygmomanometer is not available in the community.
- After discharge from hospital, heart rate, blood pressure weight and routine urinalysis is monitored in the community by either the General Practitioner or a community nursing service.
- The treatment protocol requires a weaning regimen.

Treatment Protocol

- Treatment is based on the United Kingdom Infantile Spasms Study protocol[2].
- The dose to be prescribed will be determined by the Neurologist after assessing the child.
- The usual dose is:
  - Prednisolone 10mg orally four times a day for 14 days
- If spasms continue, then on day 7 the Neurologist may increase the dose to Prednisolone 20mg orally three times a day.
- If spasms reappear between day 8 and day 14 then increase the dose to Prednisolone 20mg orally three times a day. The duration of this increased dose will be determined by the neurologist.
- Tapering of prednisolone is important and required to prevent acute adrenocortical insufficiency
- Ranitidine, omeprazole or esomeprazole should be considered to help prevent gastric ulcer that may be caused by high dose prednisolone.

Nursing Principles

After initial management in the acute setting treatment with high dose oral prednisolone will usually continue for 4-6 weeks in the community. It is important to notify the Clinical Nurse Consultant (CNC) at commencement of the therapy. The CNC will provide education, support and information to the family throughout the admission. They will also assist in organising necessary equipment, and refer to the appropriate community health care personnel prior to discharge.

Side effects of therapy and their management

1. A **rare** but potentially reversible side effect is hypertrophic cardiomyopathy which may cause unexplained tachycardia, dyspnoea, or cardiac failure. If undetected, sudden death may occur.
2. Hypertension is a serious side effect of high dose oral prednisolone.
3. Relative adrenocortical insufficiency may continue after the treatment has been ceased.
4. Electrolyte disturbances, in particular hypokalaemia.
   A full blood count (FBC) and baseline serum chemistry (electrolytes, urea and creatinine (EUC), calcium, magnesium and phosphate (CMP)) is attended prior to commencement of therapy and weekly thereafter.
5. Increased susceptibility to infection. Treatment may cause a level of immunosuppression. It is important that the parents / caregivers are advised to avoid contact with infectious persons, **in particular those with varicella**. Medical advice will be required should the child become unwell, develop a fever or come in contact with chicken pox during treatment.

6. Due to potential abnormal responses to live vaccines, **no live vaccines (e.g. MMR, MMR-V, varicella, BCG) should be given during therapy**, and for 1 month after cessation of therapy, as determined by the Neurologist. (Refer to the Immunisation protocol to be provided to the GP/community personnel).
7. General irritability, restlessness, wakefulness or changes in normal behaviour may be present throughout the course of the treatment. The parents/caregivers may need extra support and reassurance during this time.
8. Change in facial appearance, mild facial acne.
9. Increased appetite. It may be helpful to offer the infant smaller portions/volumes of formula, more frequently throughout the day.
10. Weight gain, which will resolve as treatment is weaned. Weight will be monitored weekly and should weight gain be excessive, alterations to treatment may be required.
11. Glycosuria. This will be detected on routine urinalysis and should be followed with a formal blood sugar level.
12. Urinary tract infection. This may also be detected on routine urinalysis and may need to be followed by urine microscopy.

Note: Infants do not experience all side effects and the parents/caregivers should be reassured that the side effects will almost certainly resolve once the treatment is ceased.

Adrenocortical insufficiency

Relative adrenocortical insufficiency may continue after the treatment has been ceased and a tapering course of oral prednisolone added. This may persist for several months after stopping treatment. In patients who suffer an injury or undergo surgery during or within one year after treatment, the associated stress may be managed by using rapidly acting corticosteroids. The lowest effective dose should be used, and if the dose has to be reduced it should be done gradually.

Observations

- At the commencement of the treatment manual blood pressure is measured once a day. If the blood pressure remains within documented parameters during the admission it will be measured weekly after discharge from hospital.
- Baseline weight and daily routine urinalysis during admission, followed by weekly weight and routine urinalysis once discharged.
- The following blood pressure parameters must be observed for children who receive the high dose oral prednisolone.

Age Group	Significant Hypertension	
Infant < 6 months	Systolic BP > 110mmHg	Diastolic BP > 75mmHg
Infant 6mths - 1yr	Systolic BP > 110mm Hg	Diastolic BP > 75mmHg
Children 1-5 yrs	Systolic BP > 115mmHg	Diastolic BP > 75mmHg
Children 6-10yrs	Systolic BP > 120mmHg	Diastolic BP > 80mmHg
Children 11-15yrs	Systolic BP > 125mmHg	Diastolic BP > 80mmHg
Adolescents > 15yrs	Systolic BP > 135mmHg	Diastolic BP > 85mmHg

Percentile blood pressure (mmHg) at various ages

Age in years	50th percentile		95th percentile	
	Systolic	Diastolic	Systolic	Diastolic
1	90	60	110	75
5	95	60	115	75
10	105	65	125	80
15	110	65	130	85
18	120	70	132	85

CHW Handbook 2009

Preparation for Discharge

- The CNC will assist in coordinating the discharge process and will be available for troubleshooting over the phone with the family and community health care personnel.
- When the infant is ready for discharge, the treating team will communicate with the GP/ paediatrician. The GP/paediatrician will coordinate weekly follow-up including, FBC, serum biochemistry, weight and urinalysis. Monitoring will be weekly for 1 month and then fortnightly.
- Provide to the parent/carer and community health care personnel:
  - The **high dose oral prednisolone information sheet**: http://chw.schn.health.nsw.gov.au/ou/neurology/resources/clinical_guidelines/oral_steroid_side_effects.doc
  - The **immunisation information sheet**: http://chw.schn.health.nsw.gov.au/ou/neurology/resources/clinical_guidelines/immunisation_protocol.doc
- For oral prednisolone an internal script for 7 days is provided with an external script for the continued duration of the treatment.
- The treating Neurologist may consider a referral to occupational/physiotherapist for a baseline developmental assessment. This will assist in determining the infants need for early intervention. This assessment should be considered once the child has completed the treatment.

References

1. Hockenberry MJ, Wilson D, Winkelstein M J, & Kline NE. Wong's Nursing care of infants & children, (7th Ed). Mosby, St Louis. 2003
2. Lux A I, Edwards S W, Hancock E *et al*. The United Kingdom Infantile Spasms Study comparing vigabatrin with prednisolone or tetracosactide at 14 days: a multi-centre randomised controlled trial. Lancet. 2004: 364(9447): pp.1773-8
3. Arya R, Shinnar S, Glauser TA. Corticosteroids for the treatment of infantile spasms: a systematic review. [Review] Journal of Child Neurology. 2012:27(10):1284-8.