

South Australian Paediatric Practice Guidelines

propranolol treatment for infantile haemangioma

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Note

This guideline provides advice of a general nature. This statewide guideline has been prepared to promote and facilitate standardisation and consistency of practice, using a multidisciplinary approach. The guideline is based on a review of published evidence and expert opinion.

Information in this statewide guideline is current at the time of publication.

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Health practitioners in the South Australian public health sector are expected to review specific details of each patient and professionally assess the applicability of the relevant guideline to that clinical situation.

If for good clinical reasons, a decision is made to depart from the guideline, the responsible clinician must document in the patient's medical record, the decision made, by whom, and detailed reasons for the departure from the guideline.

This statewide guideline does not address all the elements of clinical practice and assumes that the individual clinicians are responsible for discussing care with consumers in an environment that is culturally appropriate and which enables respectful confidential discussion. This includes:

- The use of interpreter services where necessary,
- Advising consumers of their choice and ensuring informed consent is obtained,
- Providing care within scope of practice, meeting all legislative requirements and maintaining standards of professional conduct, and
- Documenting all care in accordance with mandatory and local requirements

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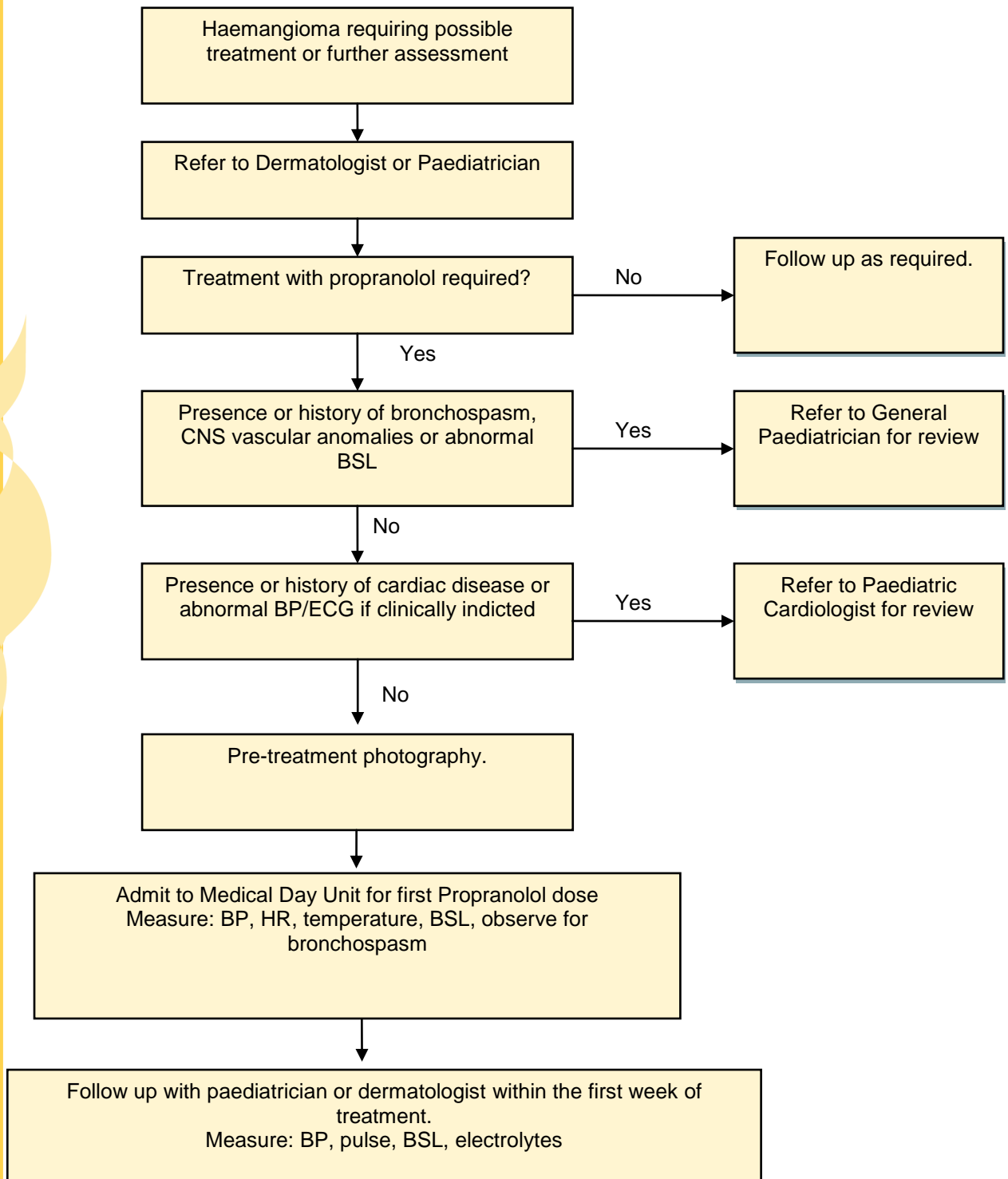
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Management flowchart for the use of Propranolol in infantile haemangiomas



Important points

- > Haemangiomas usually do not require treatment, however treatment is required if:
 - > the location is life-threatening such as airway obstruction or cardiac failure
 - > there are local complications
 - > there are cosmetic concerns especially lip, nose and ear sites
 - > there are functional risks such as potential anatomical distortion and scarring.¹
- > Patients must be reviewed by a paediatrician and dermatologist before commencing propranolol.
- > Patients must be reviewed by a paediatrician or dermatologist during the first 7 days of treatment.

Introduction

- > Infantile haemangiomas are the most common soft tissue tumour in children aged less than 1 year and occur in 4-10% of infants.
- > They are more common in female and premature infants
- > They often are not present at birth but appear in the first weeks of life.
- > In the first 3 to 6 months of life, haemangiomas may grow quickly, before entering a stabilisation phase.¹ Subsequently, haemangiomas begin to spontaneously resolve and regression is complete in 60% of 4 year olds and 76% of 7 year olds.
- > Propranolol is emerging as an effective and relatively safe treatment option. This guideline intends to standardise its use in this setting.

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Definitions and abbreviations

- > Synonyms of infantile haemangioma include: haemangioma of infancy, strawberry naevus.
- > Subgroups include congenital haemangiomas
 - RICH = rapidly involuting congenital haemangioma,
 - NICH = non-involuting congenital haemangioma
- > It is critical to distinguish infantile haemangioma from the group of birthmarks classified as vascular malformations⁴. Forms of vascular malformations include:
 - capillary ("port-wine stain"),
 - venous,
 - lymphatic
 - mixed,
 - arterio-venous
- > Vascular malformations are usually well-formed at birth, do not involute with time, and have not been shown to respond to treatment with propranolol.
- > PHACES syndrome is a rare association of infantile haemangioma,
 - P - posterior cranial fossa abnormalities
 - H - haemangioma
 - A - arterial anomalies
 - C - cardiac including aortic coarctation
 - E - eye
 - S - sternal and midline clefts
 - Due to potential vascular and cardiac involvement propranolol may have additional risks in these patients

Scope of guideline

This guideline is aimed at medical staff who are involved in the treatment of children with infantile haemangiomas. The use of Propranolol is a relatively new treatment modality and should only be started with the involvement of tertiary specialist paediatric and dermatology services. These guidelines are based on anecdotal and consensus opinion, with double-blind studies underway at a number of international locations and definitive results awaited.

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Assessment

- > The patient needs to be assessed to determine if treatment is necessary.
Treatment may be required if there are:
 - potentially life- or function- threatening problems including cardiac failure, respiratory distress or ocular compromise
 - potential anatomical distortion and scarring, especially lip, nose and ear sites
 - cosmetic considerations.
- > The presence of a history of bronchospasm, cardiac disease, CNS vascular anomalies (suspected PHACE syndrome (a syndrome involving facial haemangiomas, cardiovascular and intracranial anomalies), large cervicofacial haemangiomas) need to be excluded or further investigated before initiating treatment with propranolol.
- > All patients should be reviewed by a dermatologist and paediatrician.
- > Patients from rural and regional centres may consult with the Women's and Children's Hospital dermatology department, and sending digital photographs or a telehealth conference may be considered. Following this assessment the patient may need to transfer to a tertiary hospital for more detailed evaluation and commencement of treatment.

Management

Initial management

- > Baseline investigations
 - Blood Sugar Level (BSL)
 - Blood pressure (BP)
 - Electrocardiograph (ECG) if clinically indicated.
- > A General Paediatrician or Paediatric Cardiologist (as appropriate) should be consulted if any findings are concerning for evaluation and dosing recommendations.
- > Pre-treatment and follow-up clinical photography is mandatory to provide an objective assessment of progress and response to treatment.
- > Propranolol can be commenced at a dose of 2mg/kg/day in 2 divided doses, however the dose may be commenced at 1mg/kg/day before increasing to 2mg/kg/day in infants less than 2 months of age due to an increased risk of hypoglycaemia.
- > Oral steroids may be used concurrently with propranolol in consultation with a paediatric dermatologist.
- > ALL patients commencing propranolol should be admitted for observation to a Medical Day Unit or equivalent for administration of the first dose.

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- > Monitor heart rate, blood glucose levels, temperature and blood pressure and observe for bronchospasm hourly for 4 hours.
- > If significant abnormality occurs, cease Propranolol and consult with a paediatrician.

Ongoing management

- > An appointment with a paediatrician or dermatologist is required during the first 7 days of treatment. Monitor:
 - BP, pulse, BSL and electrolytes to exclude hyperkalaemia
- > Treatment often is required to continue up to 1 year of age, as the haemangioma may begin to proliferate after treatment ceases.
- > Propranolol should not be discontinued abruptly, and should be weaned over weeks or months.
- > All patients with haemangioma receiving propranolol should be monitored in consultation with a paediatric dermatologist who will guide further management if treatment with propranolol is ineffective.
- > Criteria for discontinuation include:
 1. adverse effects - cardiac, hypoglycaemia, behavioural (sleep / unsettled)
 2. poor response - progressive growth or severe ulceration
 3. planned - weaning of the therapy after 12 months of treatment (see below)

Weaning of therapy

During the first year, slow weaning of the dose according to weight is suggested provided response to treatment has been adequate (not increasing the initial numerical dose after prescription of 2-3mg/24 hours in divided doses)

At 1 year of age, dosage can be halved with review in 4-6 weeks. Cessation can then occur with review in 4-6 weeks.

Most haemangiomas grow little after weaning at 1 year and some have needed prolonged treatment (dose reinstatement may be required)

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