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EMIRATES DERMATOLOGY SOCIETY

Infantile Haemangiomas: A Few Challenging Cases

Dr Charlotte Goodhead

27TH WORLD CONGRESS OF DERMATOLOGY 2031
DUBAI - CANDIDATE CITY

Children and young people clinic

Our dermatology department is child friendly and we have a highly skilled team of Nurse Specialists and a Nursery Nurse to help distract our young patients

The Newcastle upon Tyne Hospitals NHS
NHS Foundation Trust

CONFLICT OF INTEREST

Infantile Haemangioma

Learning Objectives

- Diagnosis
- Management & complications
- Case examples
- Haemangioma syndromes
- Differential diagnosis

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Infantile haemangiomas (IHs)

Relatively common up to 5% of children

Girls 3:1

80% head and neck

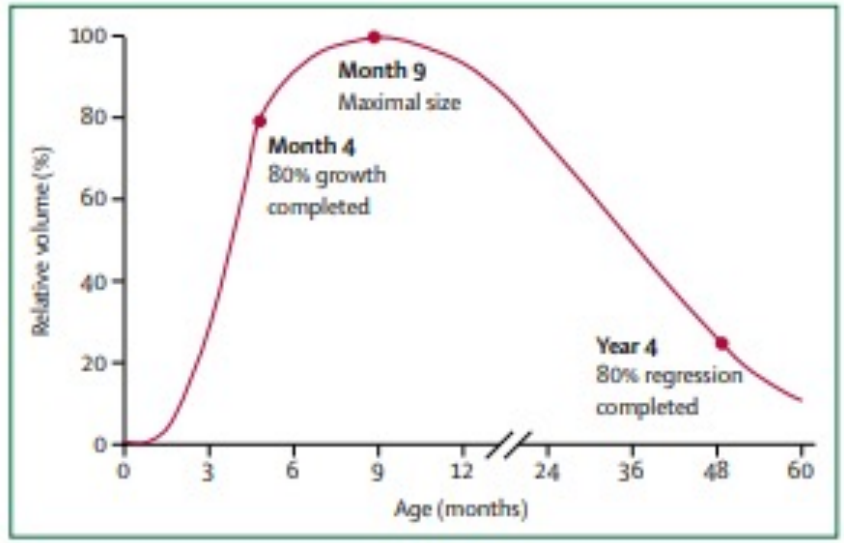
Increased frequency seen in

- Low birth weight
- Prematurity
- Multiple pregnancy (Kowalska 2021)

Complications

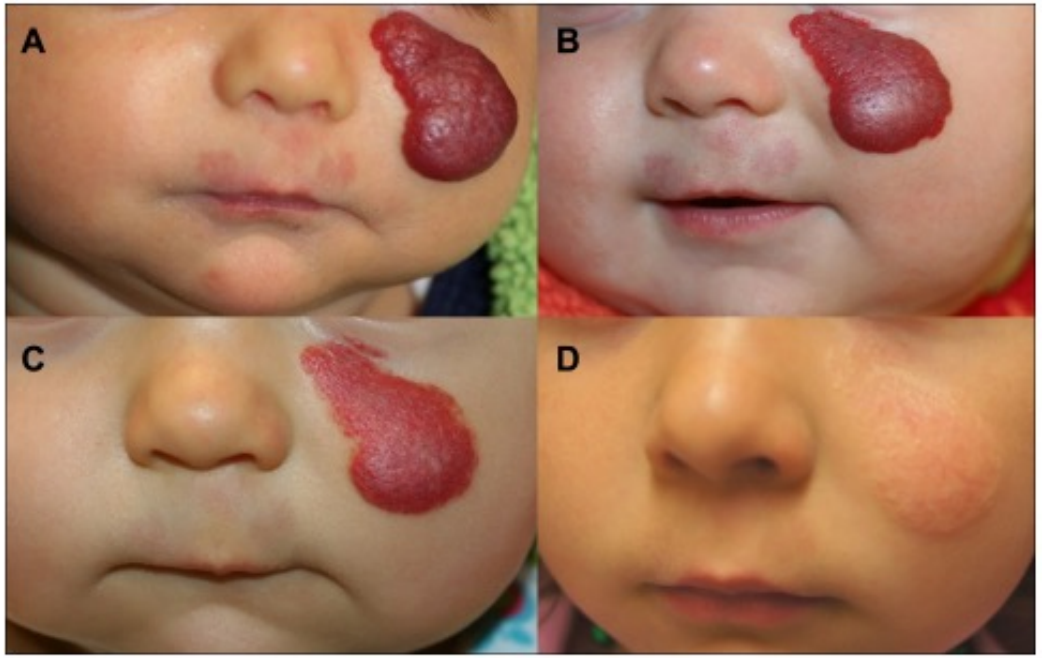
- Bleeding / Ulceration – 15%
- Functional impairment – vision, feeding, breathing, ears
- Psychosocial and disfigurement

Phases of infantile haemangioma



(Léauté-Labrèze 2017)

1. **Rapid proliferation**
   - First 3 months (especially wks 5.5 - 7.5)
   - 80% of growth achieved by 3 months
   - Maximum size by around 9 months
2. **Plateau**
3. **Slow involution**
   - Regression by age 4 years in 90% cases



(Rosenblatt 2012)

Localised

Well-defined focal lesions
Appearing to arise from a central point

Segmental

IH involving an anatomic region that is often aque-like and often measuring at >5 cm in diameter

Indeterminate (undetermined)

Neither clearly localized or segmental (often called partial segmental)

Multifocal

Multiple discrete IHs at disparate sites

(Krowchuck 2019)

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GUIDELINE

BJD
British Journal of Dermatology

Oral propranolol in the treatment of proliferating infantile haemangiomas: British Society for Paediatric Dermatology consensus guidelines*

L. Solman,[1] M. Glover,[1] P.E. Beattie,[2] H. Buckley,[3] S. Clark,[4] J.E. Gach,[5] A. Giardini,[6] I. Helbling,[7] R.J. Hewitt,[8] B. Laguda,[9] S.M. Langan,[10] A.E. Martinez,[1] R. Murphy,[11] L. Proudfoot,[12] J. Ravenscroft,[13] H. Shahidullah,[14] L. Shaw,[1] S.B. Syed,[1] L. Wells[13] and C. Flohr[15]

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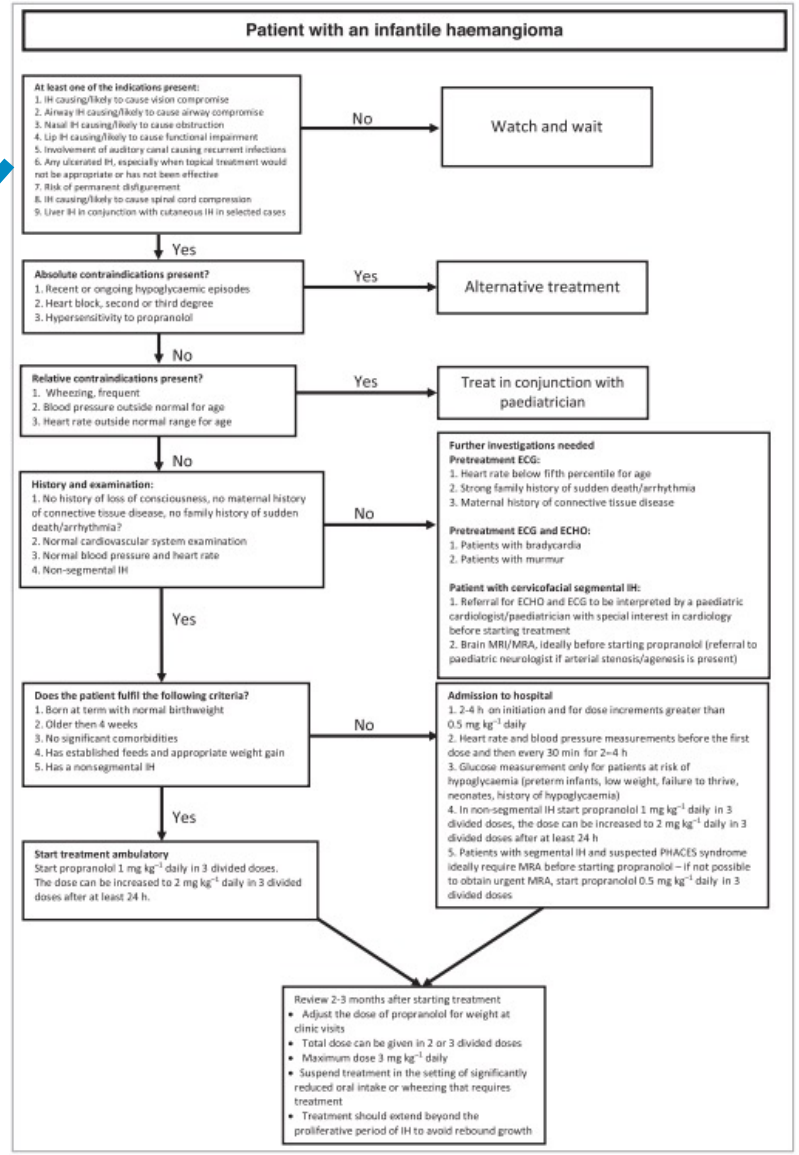
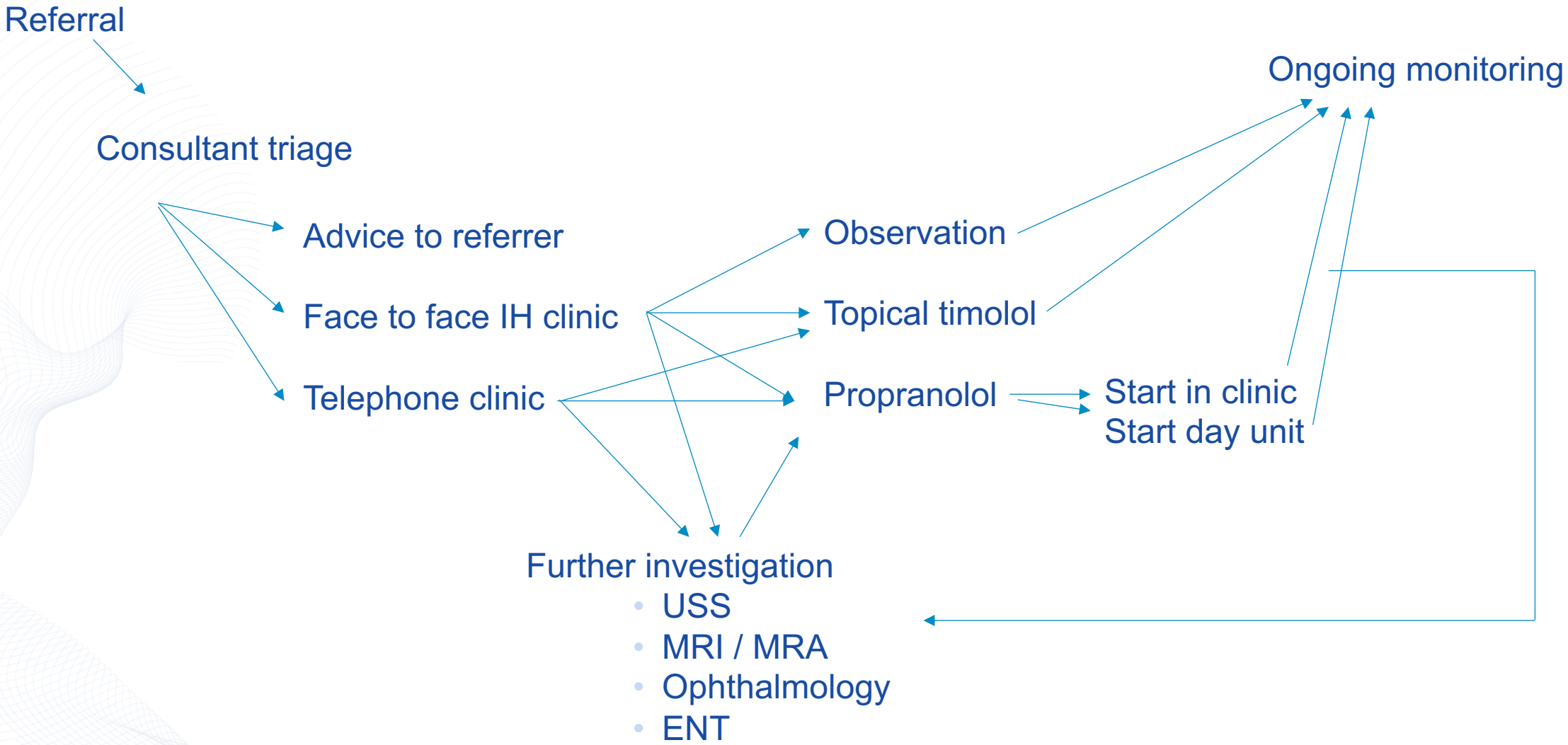
At least one of the indications present:

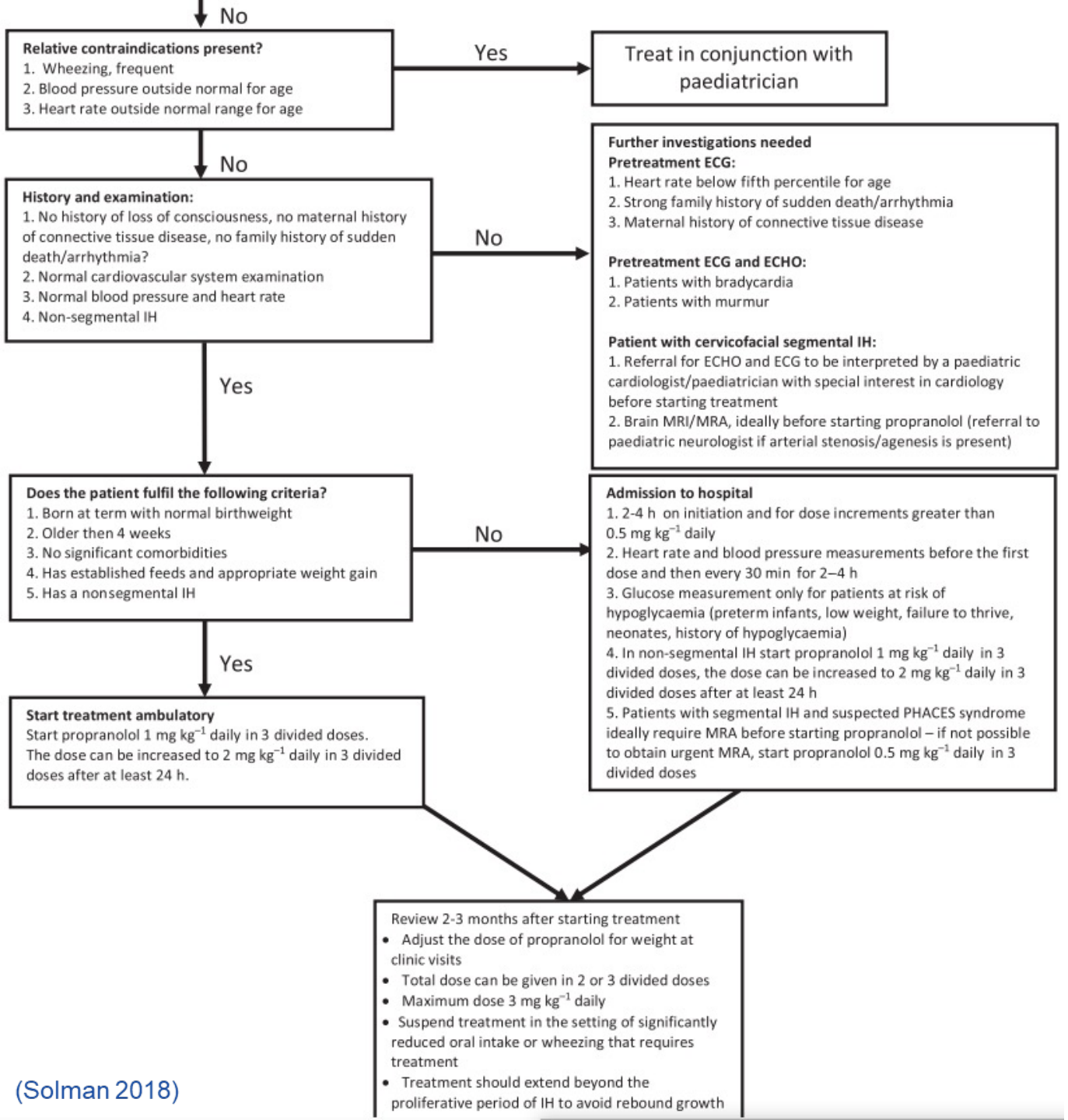
1. IH causing/likely to cause vision compromise
2. Airway IH causing/likely to cause airway compromise
3. Nasal IH causing/likely to cause obstruction
4. Lip IH causing/likely to cause functional impairment
5. Involvement of auditory canal causing recurrent infections
6. Any ulcerated IH, especially when topical treatment would not be appropriate or has not been effective
7. Risk of permanent disfigurement
8. IH causing/likely to cause spinal cord compression
9. Liver IH in conjunction with cutaneous IH in selected cases



(Solman 2018)

Management – local pathway





(Solman 2018)

- Propranolol initiated by paediatrician urgently on day unit as needed
- Can start ambulatory for those suitable – 0.5mg/kg BD increasing after 7 days to 1mg/kg BD
- 50mg/5ml solution rather than 5mg/5ml
- May see more often than 2-3 monthly to titrate dose if large, ulcerated or of concern

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Rapid growth

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Presented aged 3 weeks
Enlarging rapidly despite propranolol
Vomiting problematic

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Propranolol until 18months
Ongoing follow up with plastics

A late presentation

3.5 months old
Twin born 35 weeks

Propranolol for 1 month elsewhere
Still growing
Amblyopia and squint

Dose escalated to 3mg/kg
Admitted - prednisolone considered

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5 months later
3mg/kg propranolol

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Stopped propranolol 19months

No rebound

Aged 3 months vs 4 years

Propranolol vs Atenolol

Twin, born 31 weeks
2 months old
IH glabellum grown rapidly
IH back and buttock also

Ophthalmology review

USS and MRI – no intracranial extension

Propranolol 2mg/kg

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Propranolol caused general malaise, itching and diarrhoea
IH larger at 6 months

Changed to atenolol 1mg/kg
By 10 months improvement achieved

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PHACE syndrome

Posterior fossa brain malformation

Haemangioma facial segmental

Arterial cerebrovascular anomalies

Cardiac abnormalities or coarctation of the aorta

Eye and endocrine anomalies

LUMBAR syndrome

Lower body infantile haemangioma

Urogenital anomalies

Malformations of spinal cord

Bony defects

Ano-rectal malformations / arterial anomalies

Renal anomalies

PELVIS

Perineal haemangioma

External genitalia malformations

Lipoma myelomeningocele

Vesico-renal abnormalities

Imperforate anus

SACRAL

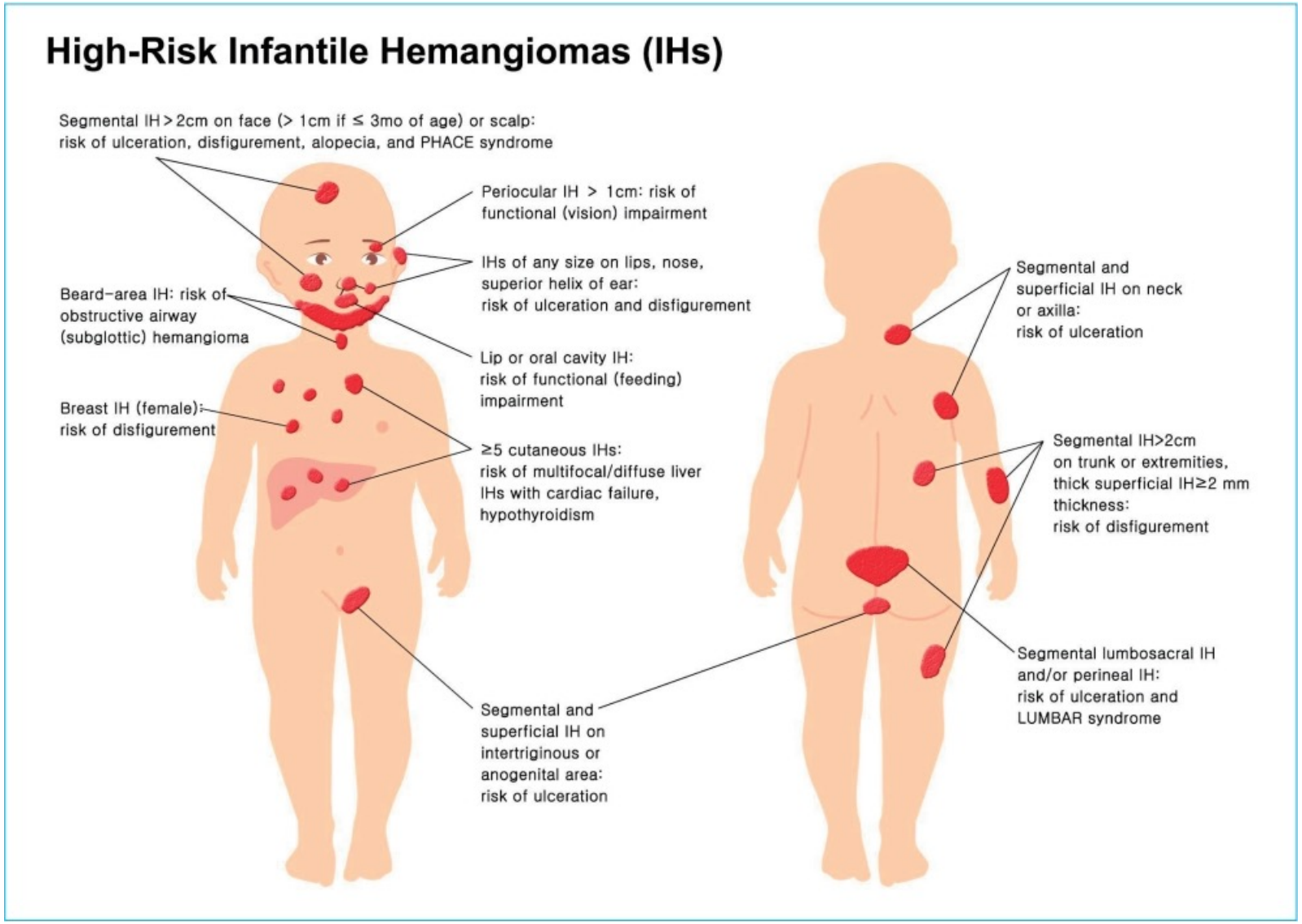
Spinal dysraphism

Ano-genital anomalies

Cutaneous anomalies

Renal and urologic anomalies

Angioma of lumbar sacral area



(Jung 2021)

PHACE

Major

- **Arterial anomalies**
  - Dysplasia, stenosis, occlusion, aberrant origin, embryonic vessels
- **Structural Brain**
  - Posterior fossa, Dandy Walker
- **Cardiovascular**
  - Aortic arch anomalies
  - Coarctation, dysplasia, aneurysm
- **Ocular**
- Posterior segment anomalies
  - Persistent hyperplastic vitreous
  - Fetal vasculature
  - Optic nerve hypoplasia
  - Peripapillary staphyloma
- **Ventral/midline**
  - Sternal pit
  - Sternal cleft,
  - Supraumbilical raphe

Minor

- **Arterial anomalies**
  - Aneurysm
- **Structural Brain**
  - Midline anomalies
  - Cortical development
- **Cardiovascular**
  - VSD
  - Right aortic arch
- **Ocular**
- Anterior segment anomalies
  - Micropthalmia
  - Coloboma
  - Cataracts
- **Ventral/midline**
  - Ectopic thyroid
  - Hypopititartism
  - Midline sternal papule

Definite PHACE

- Hemangioma>5cm in diameter of the head including scalp PLUS 1 major criteria or 2 minor criteria
- Hemangioma of the neck, upper trunk or trunk and proximal upper extremity PLUS 2 major criteria

Possible PHACE

- Hemangioma of the neck, trunk and proximal upper extremity PLUS 1 major or 2 minor
- No hemangioma PLUS 2 major criteria

(Garzon 2016)

INVESTGATIONS:

- Full examination
- Cardiology assessment
- MRI / MRA
- Ophthalmology review

Table II. Diagnostic Criteria for LUMBAR syndrome

Diagnostic criteria for LUMBAR syndrome Requires a segmental infantile hemangioma of the lumbosacral, sacrococcygeal and/or pelvic cutaneous regions* plus 1 additional criterion

Organ system	Criteria
Urogenital	• Differences in sexual development† or urogenital sinus anomalies • Other anomalies of the external genitalia ▪ Including malpositioned, bifid, atrophic, incomplete, absent, asymmetric, hypertrophied or duplicate genitalia • Uterine duplication (uterine didelphys) or vaginal duplication‡ • Bladder exstrophy/epispadias complex

Spinal Cord Malformations	• Lumbosacral spinal dysraphism/tethered cord§
	  ▪ Abnormal filum terminale in association with tethered cord¶
	  ▪ Intraspinal lipomas, intraspinal hemangiomas, myelocystocele, congenital dermal sinus tract
	• Syringomyelia/syrinx**

Bony	• Dysplasia, hypoplasia, dysgenesis, agenesis, or dissociation of the sacral or coccygeal spine
Anorectal	• Anorectal malformations ▪ Including perineal, rectourethral, recto-bladder neck, rectovaginal, or vestibular congenital fistulas • Anal or rectal stenosis • Rectal atresia • Cloaca or cloacal exstrophy
Arterial	• Aberrant origin or course, dysplasia or hypoplasia, aneurysm, stenosis, or occlusion of the aortic, renal, mesenteric, iliac, femoral, popliteal, tibial, or peroneal arteries
Renal	• Renal agenesis/solitary kidney • Renal ectopia and fusion anomalies ▪ Including pelvic kidney, horseshoe kidney, crossed-fused ectopia, or other renal malpositions

(Metry 2024)

INVESTIGATIONS:

- Full examination
- Renal USS
- MRI / MRA

Segmental distribution

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Pale, flat telangiectatic at birth

LSCS at 39 weeks

16 days old became raised

Affected ability to open left eye

Referred to ophthalmology ?PWS

Segmental or PHACE?

Seen aged 1 month
1 episode bleeding

Propranolol 2mg/kg

Cardiac assessment/
echo – normal

Increased to 2.5mg/kg at
3 months old

MRI head normal

Increased to 3mg/kg at 4
months old

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38+6 LSCS

10 days old – developed IH

Right scalp, arm, hand axilla chest

Propranolol 0.5mg/kg as becoming thickened

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Echo and ECG normal

MRI at 10 months: congenital absence of right internal carotid

Left posterior cerebral artery unusual shape but normal brain

Stopped propranolol 16 months

Residual speckling only
Transient poor growth

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Differential diagnosis

Panel 1: Main differential diagnoses of IH

Present at or soon after birth

- Vascular tumour or anomaly
- Congenital haemangioma: rapidly, partially, or non-involuting type
- Kaposiform haemangioendothelioma or tufted angioma
- Capillary malformation (port-wine stain)
- Macrocystic lymphatic malformation
- Venous anomaly
- Others: myofibromatosis, dermoid cyst, teratoma, sarcoma (fibrosarcoma), neuroblastoma, leukaemia (so-called blueberry muffin baby)

Developed after birth

- Vascular tumour or anomaly
- Pyogenic granuloma
- Macrocystic lymphatic malformation
- Glomuvenous and venous anomalies
- Kaposiform haemangioendothelioma
- Malignant tumours (sarcoma, lymphoma, cutaneous localisation of neuroblastoma, or leukaemia)
- Others: haematoma, benign tumours (pilomatrixoma, Spitz naevus, myofibromatosis, neurofibroma, eosinophilic granuloma, myxoma, lipoblastoma, siloblastoma)

(Solman 2018)

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