St George's University Hospitals NHS
NHS Foundation Trust

All that itches and changes colour

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NHS Foundation Trust

Paediatric Dermatology at St Georges

- Weekly paediatric clinic (Friday afternoon)
- Weekly adolescent clinic (Tuesday morning)
- Nurse led eczema education (Friday afternoon)

- On call Dermatology Service

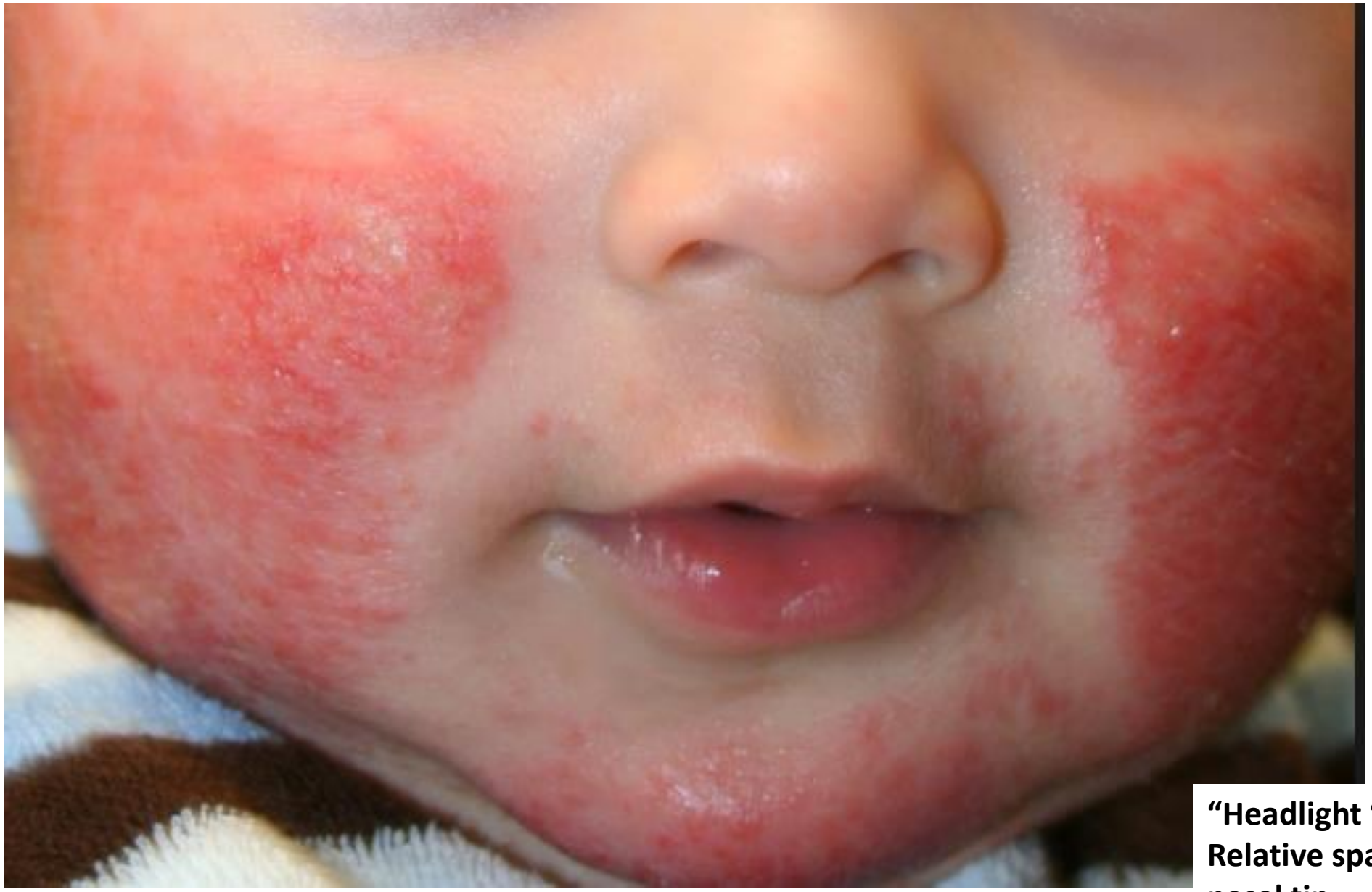
Aim

- To discuss approaches and strategies to assess and treat children with atopic dermatitis – latest evidence
- Pathway for management of AD in children
- To review the classification of infantile haemangiomas
- Which patients with haemangiomas need to be referred and current treatment options

To start with all that itches…..

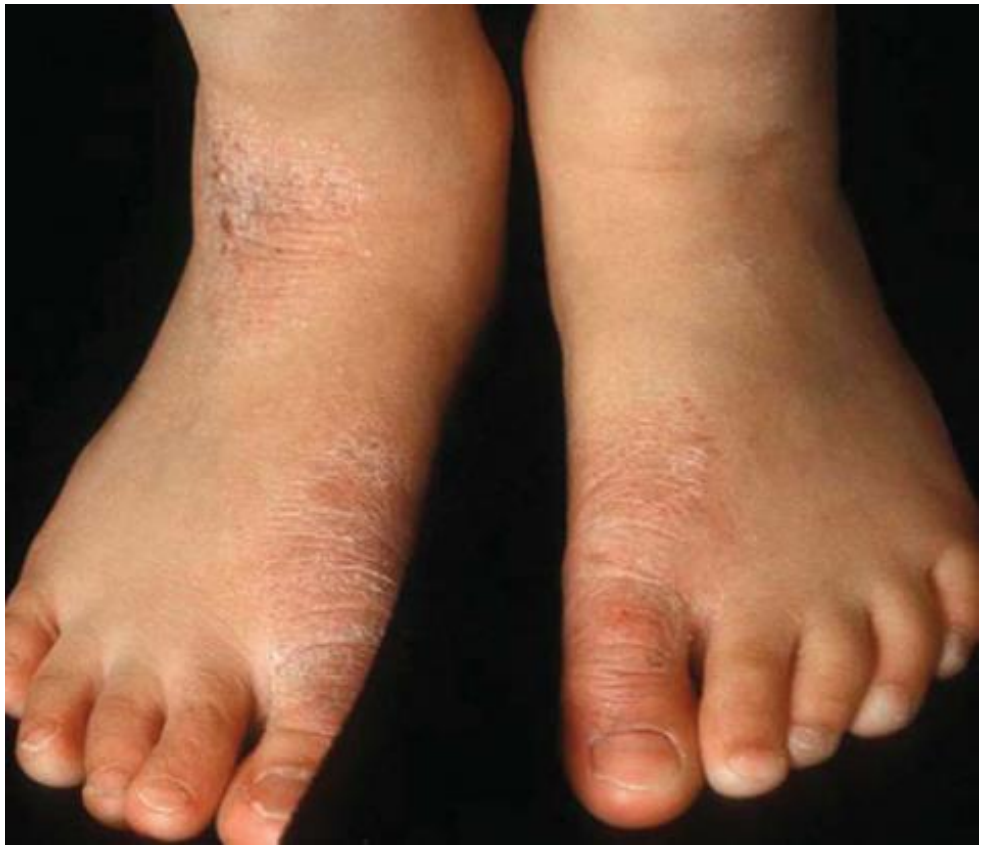
Atopic Dermatitis

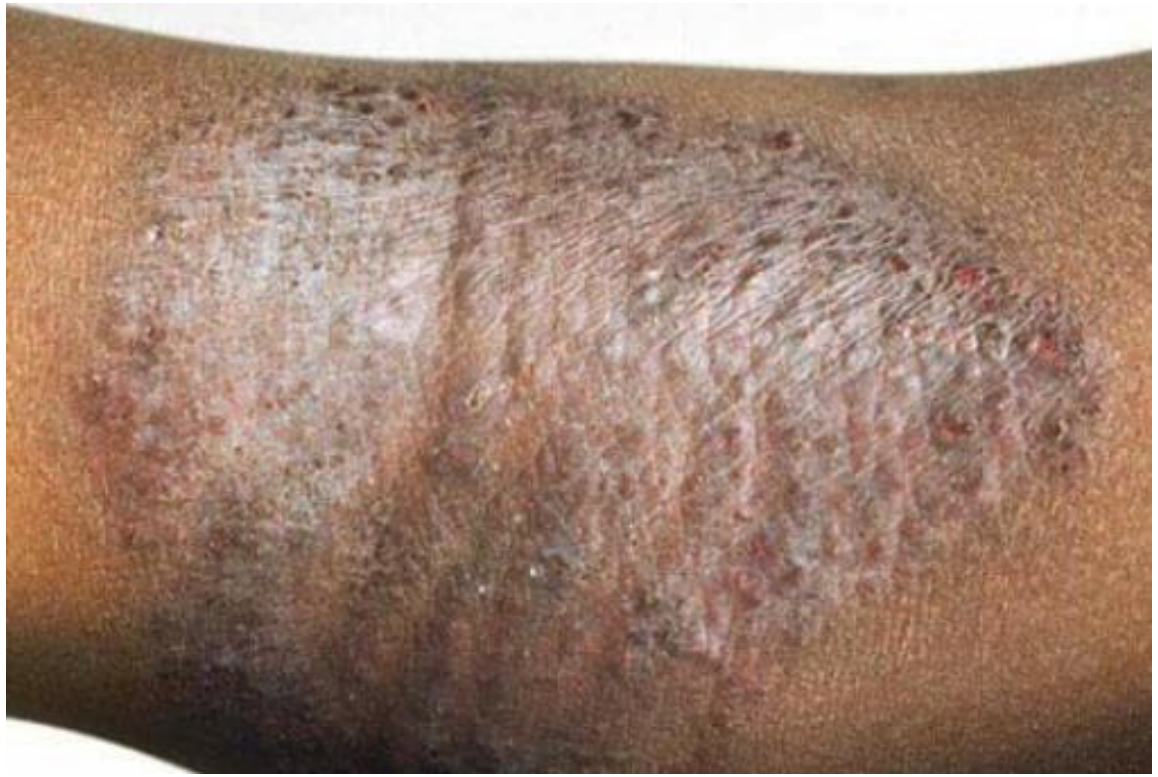
- One of the most common skin disorders seen in children
- Prevalence of 10-13%
- **Essential feature** – itch in past 12 months
- **Plus three of the following**
  - History of dry skin over the last year
  - Personal history of atopic disorder (or family history in first degree relative if <4years)
  - History of skin crease involvement
  - Visible flexural dermatitis (if child <4 years include cheek, forehead and extensor surfaces of limbs)



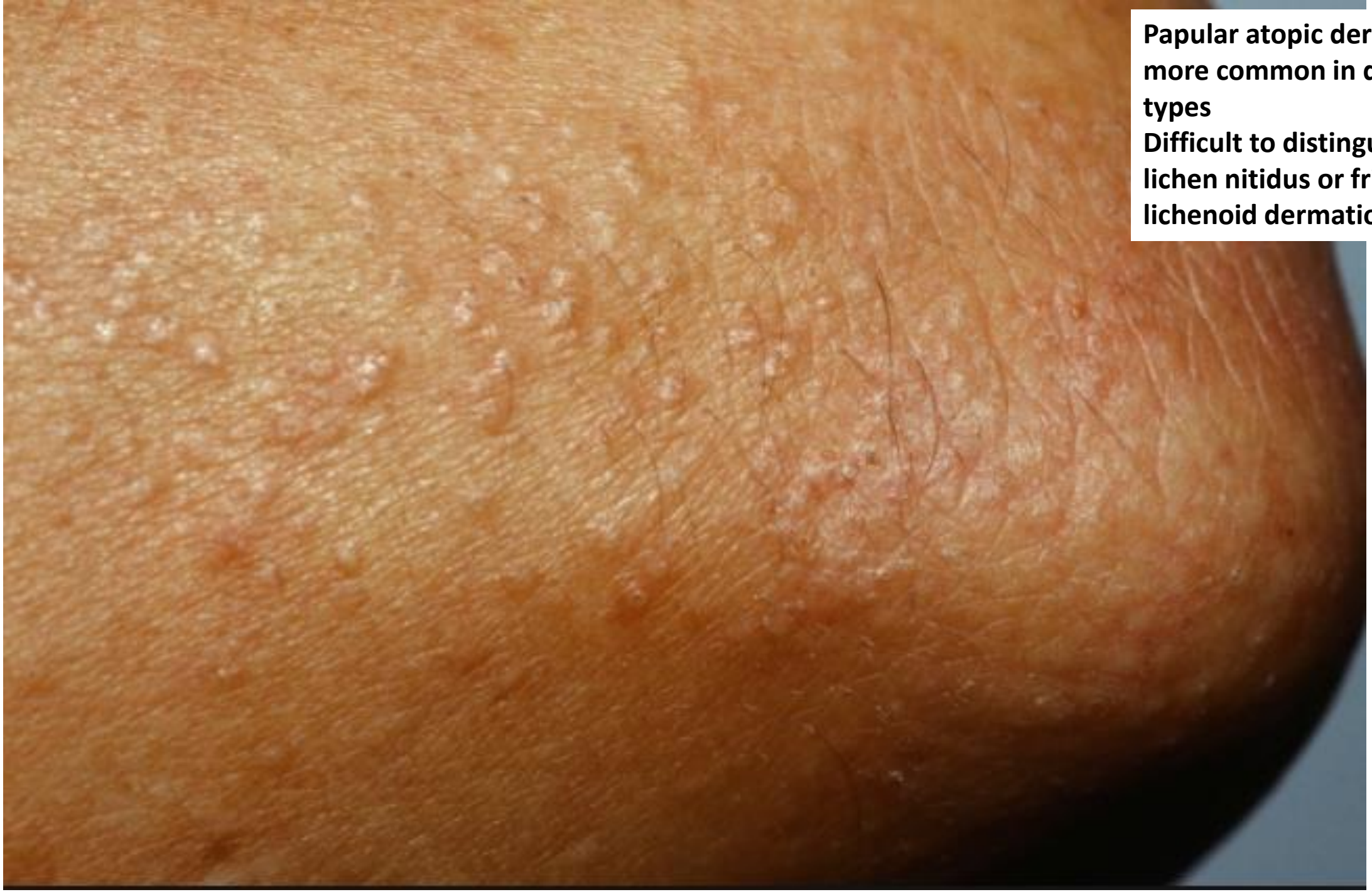
“Headlight “ sign
Relative sparing of midface, nasal tip
and immediate perioral area

Involvement of extensor surfaces during infant phase Crawling





Lichenification in classical areas after aged 2 years



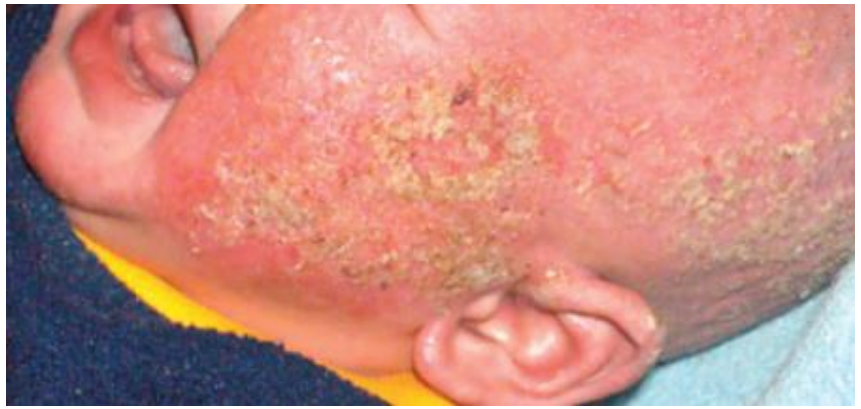
Papular atopic dermatitis more common in darker skin types
Difficult to distinguish from lichen nitidus or frictional lichenoid dermatiosis

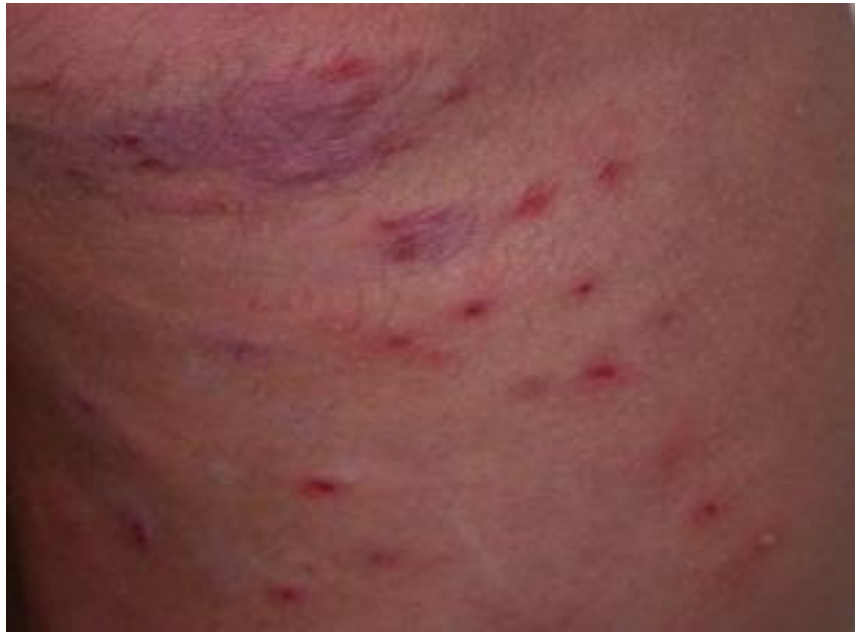
Focused history for an eczema consult

- **Age on onset**
- **Triggers**
- **Family history of atopy**
- **Quality of life assessment (sleep disturbance/school attendance/poor concentration)**
- **Recent infections**
- **Treatment history**

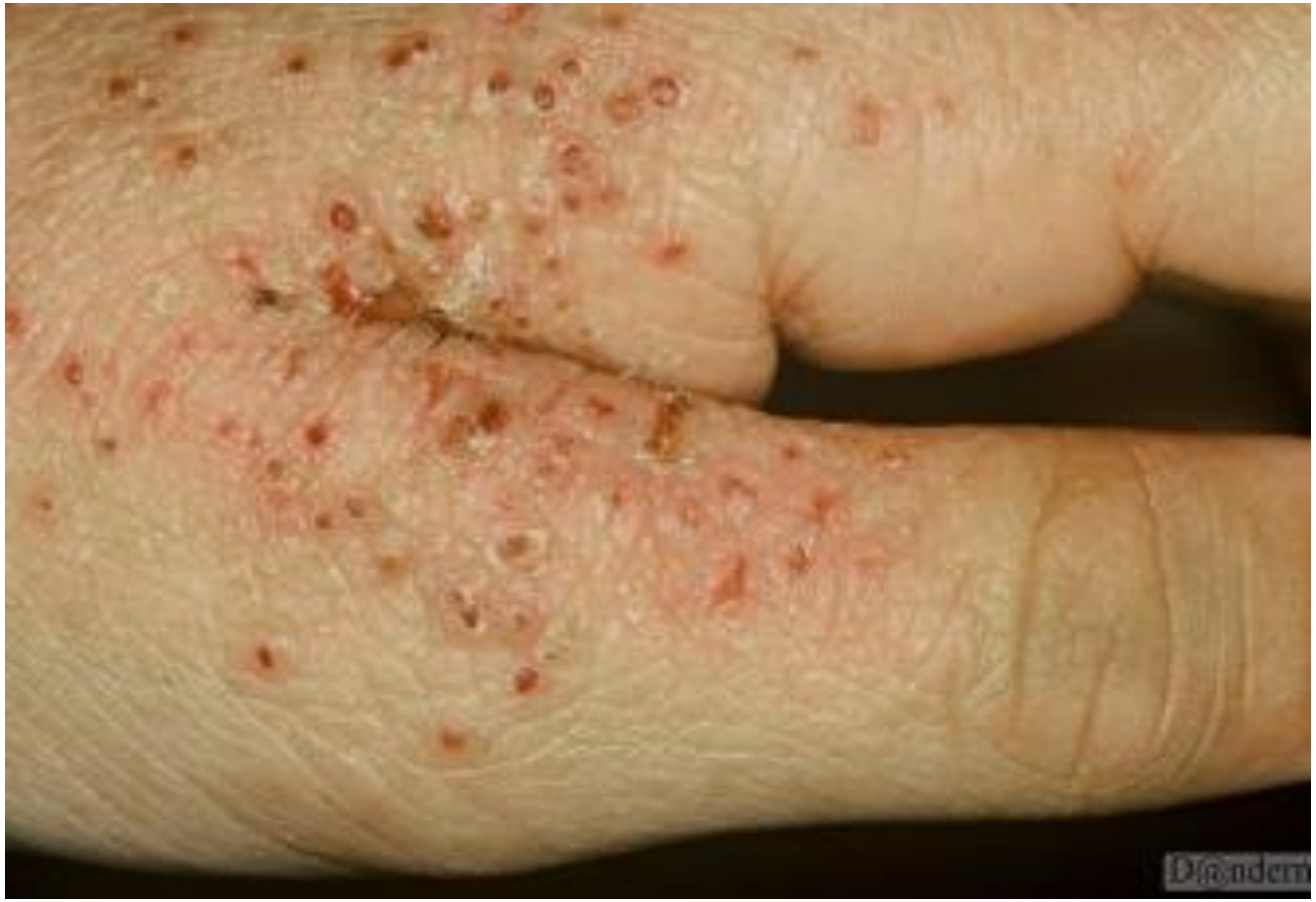
Infectious complications - bacterial

- Staph aureus is found in up to 90% of patients with AD (76% from uninvolved skin and 50-60% from nares)
- Although secondary infection is usually from staph aureus, 16% show Strep and 14% mixed orgs → therefore always swab
- Consider if
  - Pustules
  - Exudate
  - Crusting
  - Flare of chronic AD or fails to respond to appropriate therapy



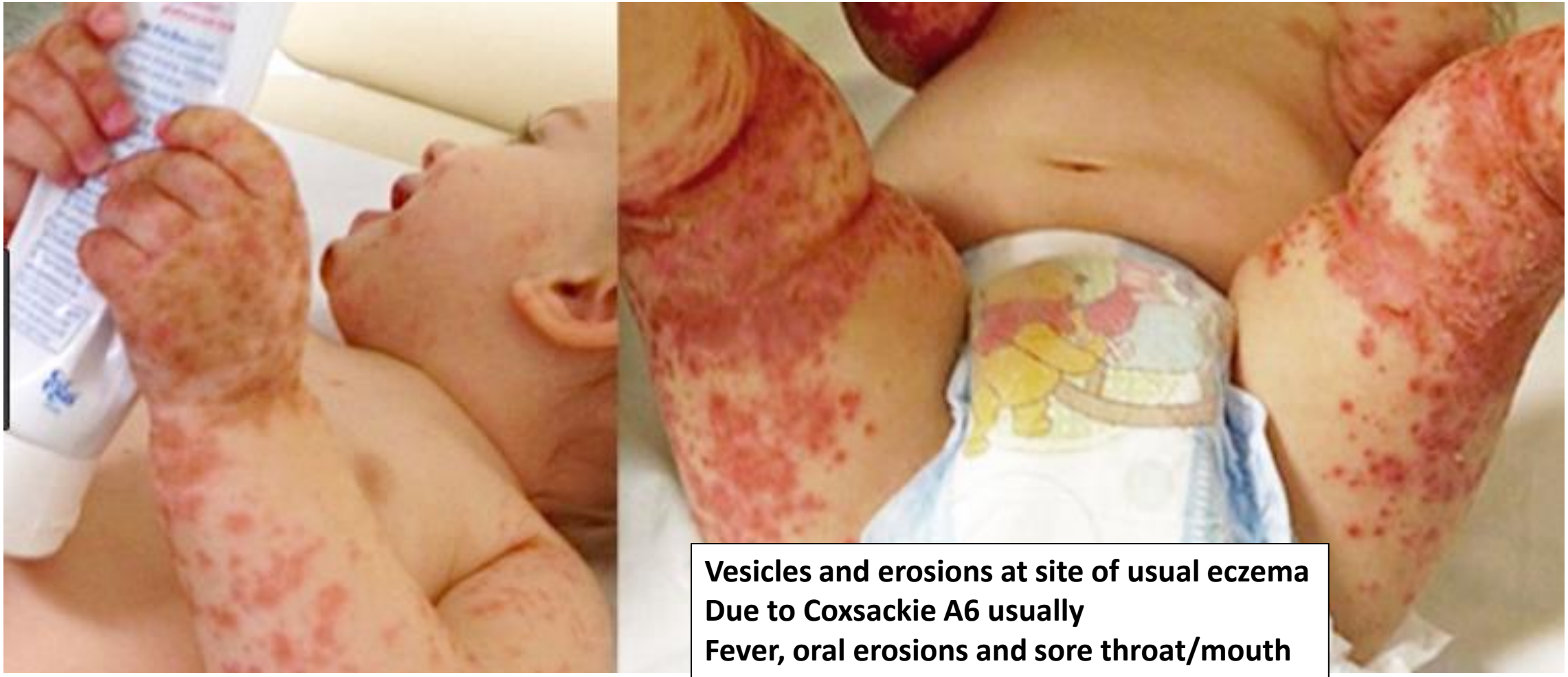


Infectious complications – eczema herpeticum



- Explosive vesiculopustular eruption due to HSV
- Hospitiliastion may be necessary esp if under 1 or systemic symptoms
- Early administration of acyclovir
- Systemic antibiotics usually needed

Infectious complications – eczema coxsackium



Vesicles and erosions at site of usual eczema
Due to Coxsackie A6 usually
Fever, oral erosions and sore throat/mouth
Lesions usually clear 12 days – 1month

Prognosis and impact on QOL

- Previous studies suggested eczema tends to clear in up to 70% by puberty, but now more than 80% of children with mild/moderate AD have recurrent symptoms requiring meds into early 20s
- Sleep disturbance in up to 60% of children
- Strong association with ADHD
- Increased odds ratio of depression, anxiety and learning delay

J Invest Dermatol. 2014 Jul;134(7):1847-1854. doi: 10.1038/jid.2014.70. Epub 2014 Feb 4.

Suicidal ideation, mental health problems, and social function in adolescents with eczema: a population-based study.

Halvorsen JA[1], Lien L[2], Dalgard F[3], Bjertness E[4], Stern RS[5].

Management

- Patient and parent education
- Avoidance of irritants and allergic triggers
  - Products marked as “sensitive/organic/gentle” were found to have 132% more reference allergens than own brand products
- Main issue is **undertreatment and anxiety regarding topical steroid use**
- Written skin care plans have been shown to improve adherence

Emollients

- Ointments > creams (depending on tolerance)
- Oils not able to penetrate skin as well as oil/water mixtures
- Avoid olive oil detrimental to skin barrier recovery
- Soap substitutes essential
- Limit bathing to 10 minutes a day
- Bleach baths (anti-inflammatory and anti-bacterial)
- Environmental factors (temperature, avoid sweating, swimming is fine but need prep, avoid overdressing, soft cotton clothing)

Assessment and modification of triggers

- Food allergy testing for milk, egg, peanut, wheat and soy if **< 5 years with moderate/severe AD who have persistent AD despite optimum mx or if clear history of food related exacerbations**
- Aeroallergens : house dust mite, grass, animal dander, moulds – particularly for older children with exposed site eczema

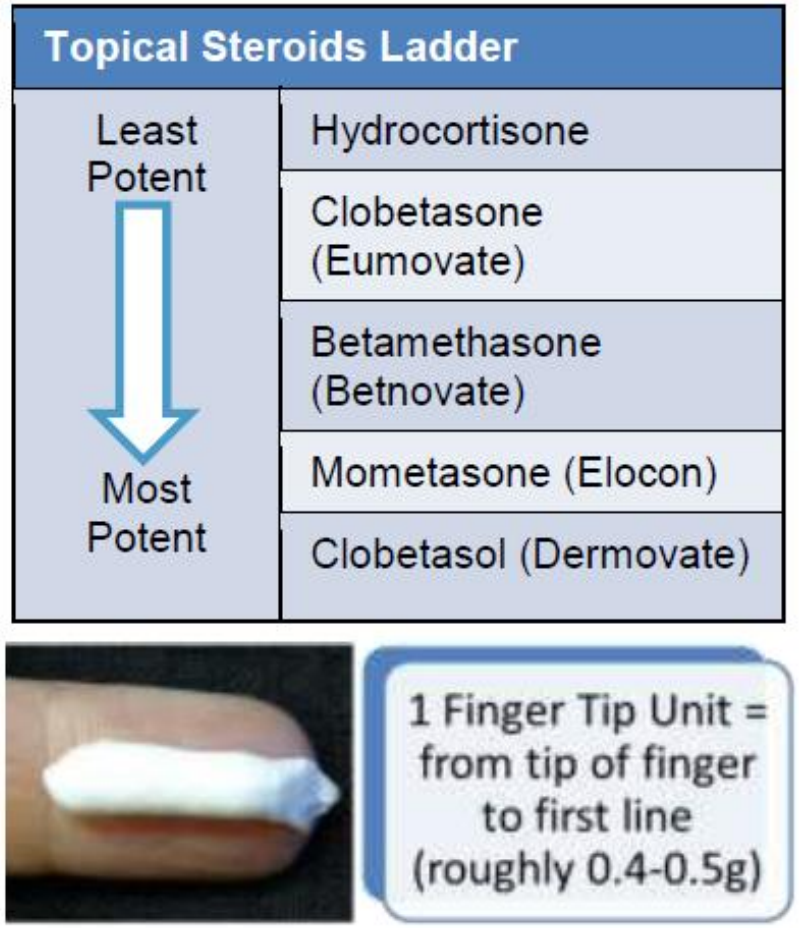
Topical steroids – most pressing issue is undertreatment

- **81% of families with AD have steroid phobia and 36% admit non-adherence as a result**
- Evidence to show safety of week-end treatment with either TCS or calcineurin inhibitor in children
- Aim to prescribe ointments as more effective (although creams may be preferred during summer months as less sweat retention, can contain additives)
- Topical calcineurin inhibitors eg Protopic 0.03% and Elidel licensed in over 2 years – only confirmed safety issue is burning and itch on application, apply at night
- New trials of phosphodiesterase 4 inhibitors are currently ongoing

Eczema treatment guideline

Mild eczema	Moderate eczema	Severe eczema
For acute flares apply a topical steroid (eg Hyrdocortisone) daily for at least two weeks Consider maintenance treatment with twice weekly consecutive days applications	For acute flares apply a topical moderately potent steroid (eg Eumavate,) daily for at least two weeks Consider topical calcineurin inhibitors eg Protopic ointment (can contact via KINESIS for specialist advice if needed) Consider maintenance treatment with twice weekly consecutive days applications	For acute flares apply a topical potent steroid (eg Elocon) daily for at least two weeks Consider topical calcineurin inhibitors eg Protopic ointment(can contact via KINESIS for specialist advice if needed) Consider maintenance treatment with twice weekly consecutive days applications

How much is too much ?



AGE	FACE & NECK	1 ARM & HAND	1 LEG & FOOT	TRUNK (FRONT)	TRUNK (BACK) INCLUDING BUTTOCKS
3–6 MONTHS	1	1	1.5	1	1.5
1–2 YEARS	1.5	1.5	2	2	3
3–5 YEARS	1.5	2	3	3	3.5
6–10 YEARS	2	2.5	4.5	3.5	5
10+ –ADULTS	2.5	4	8	7	8

- Using superpotent twice daily for longer than 6-8 weeks (less if delicate area)
- Over treatment is rarely an issue
- Written guidance is very important

Whats the role of antihistamines in eczema

- No value
- Can alleviate allergic rhinitis and may be helpful when facial/periorbital eczema is an issue
- Essentially this is purely for sedation – topical treatments are the mainstay

Role for treatment of secondary cutaneous infections

- In US bleach baths (dilute Milton baths twice a week) standard care for decreasing moderate to severe AD
- Antiseptic washes such as Dermol range of soap substitutes if recurrent infections

When to refer ?

Paediatric Eczema Referral Form

Pre-referral criteria

1. The patient is using a daily emollient (min. 250g/week) Yes ☐

2. The patient is using a soap substitute Yes ☐

3. The patient has been tried on a topical potent steroid (eg Betnovate ointment, Elocon ointment) for at least 2 weeks to the body or a moderately potent steroid (eg Eumavate ointment) for 7 days to the face Yes ☐

Routine referral to Dermatology may be appropriate if all of the above have been done.

NICE guidance for referral

1. Diagnosis uncertain
2. Management not controlled (>1 flare month)
3. Atopic eczema on face not controlled with standard treatments and need advice considering TCI
4. Advice on wet wraps or bandaging
5. Possible contact allergy
6. Significant social or psychological problems
7. Eczema associated with severe or recurrent skin infections

Management of severe atopic dermatitis

- TL01 narrow band UVB phototherapy
  - Moderate improvement in 89% of chidren; complete clearance in 40% over 3 months
  - School attendance major factor
- Systemic steroids
  - significant rebound after stopping, side effects
- Methotrexate
  - Not enough large trials in children but effective in 75%, minimal adverse effects
- Ciclosporin
  - Most rapid mode of action but higher side effects and limitation of use
- Mycofenalate mofetil
- Biologic (Dupilumab, anti IL-4 antibody, trials in adults)

And now to things that change colour…..

Vascular birthmarks

Cellular proliferation

Errors in vascular morphogenesis; dysplastic vessels

Vascular tumours

- **Infantile haemangioma**
- **Congenital haemangioma(RICH/NICH)**
- **Kaposiform haemangioendothelioma**
- **Tufted angioma**
- **Pyogenic granuloma**

Vascular malformations

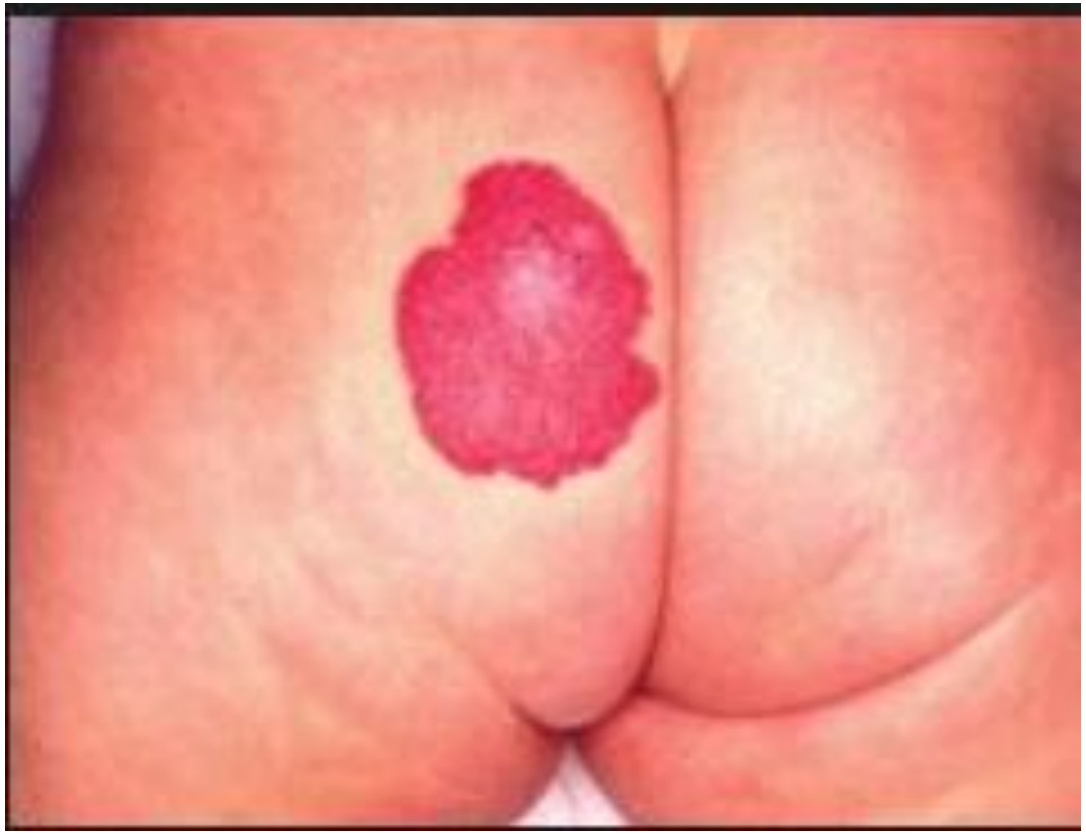
- **Capillary malformation**
- **Venous malformation**
- **Lymphatic malformations**
- **Arteriovenous malformation**

Which term is the correct one ?

- Strawberry naevus
- Strawberry haemangioma
- Cavernous haemangioma
- Infantile haemangioma
- Capillary haemangioma
- Infantile capillary haemangioma

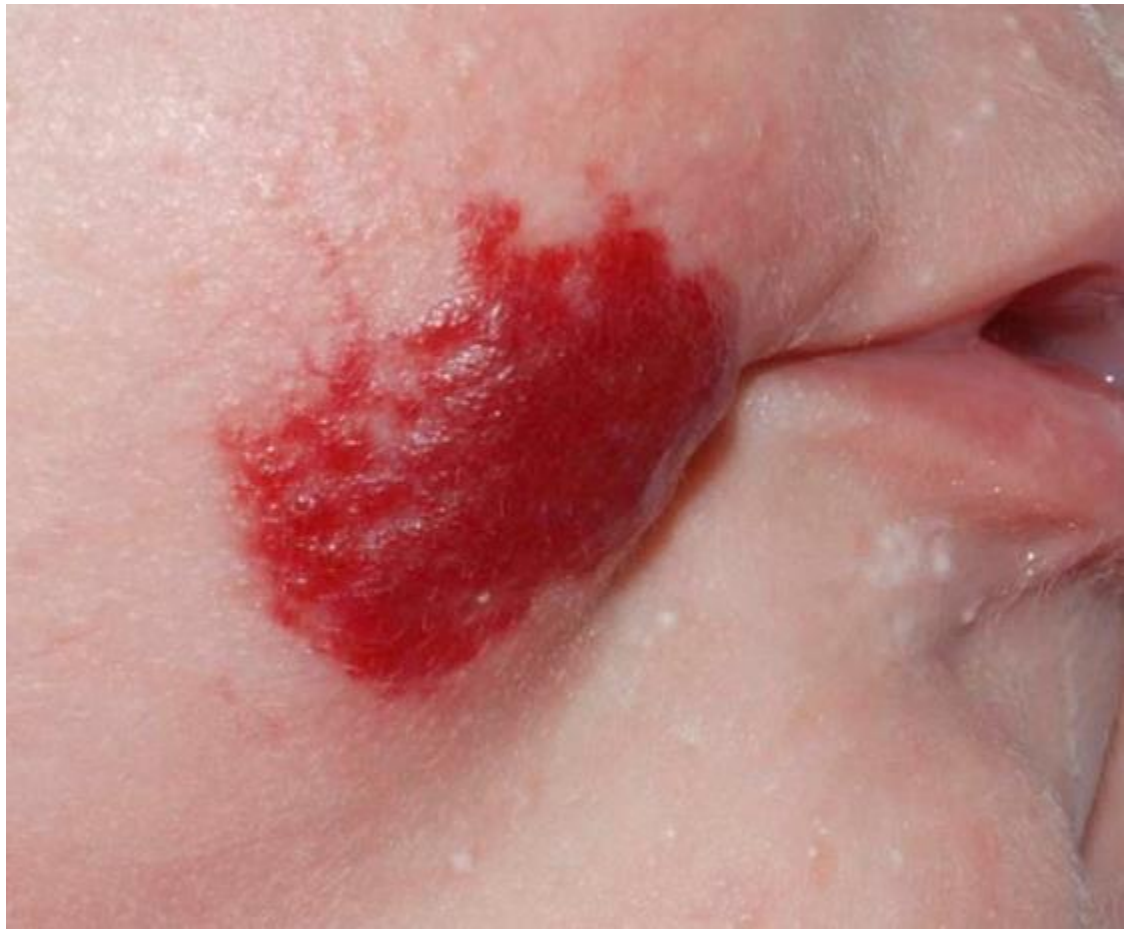
Infantile Haemangioma

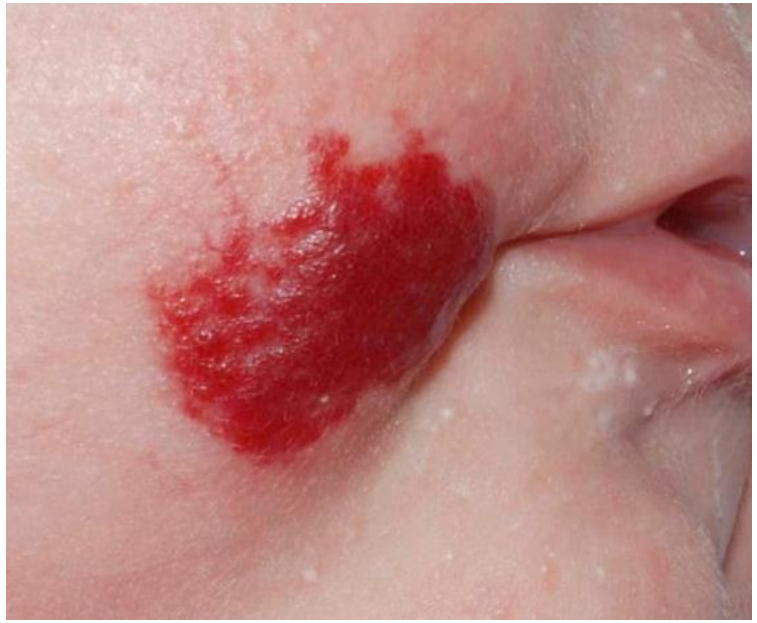
- Common
- Develop in 4-5% of infants
- Caucasian
- F>M
- More frequent in pre-term (15% of BW 1- 1.5kg), older mothers, multiple gestation, low birth weight



Clinical Features

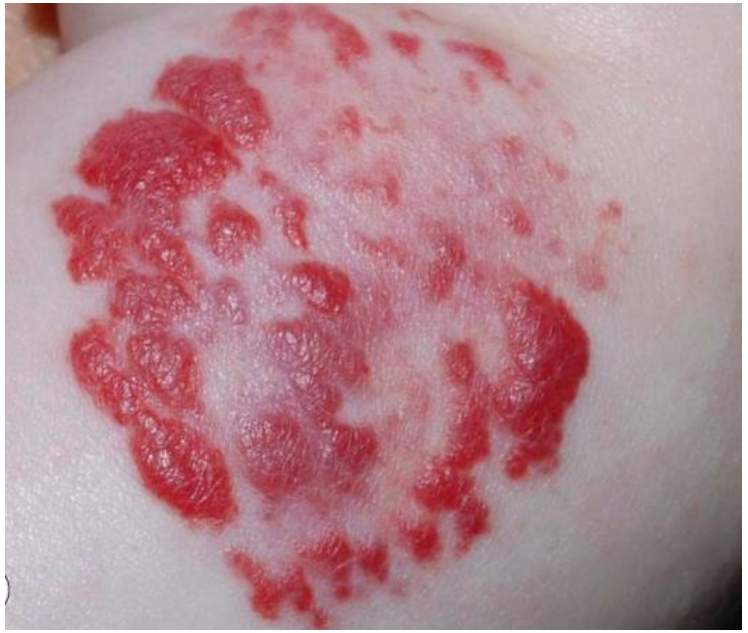
- Usually become apparent in first few weeks
- Congenital lesions may be precursor lesions
- 50% on head and neck
- Most common are superficial (50-60%), mixed (25-35%) and deep (15%)

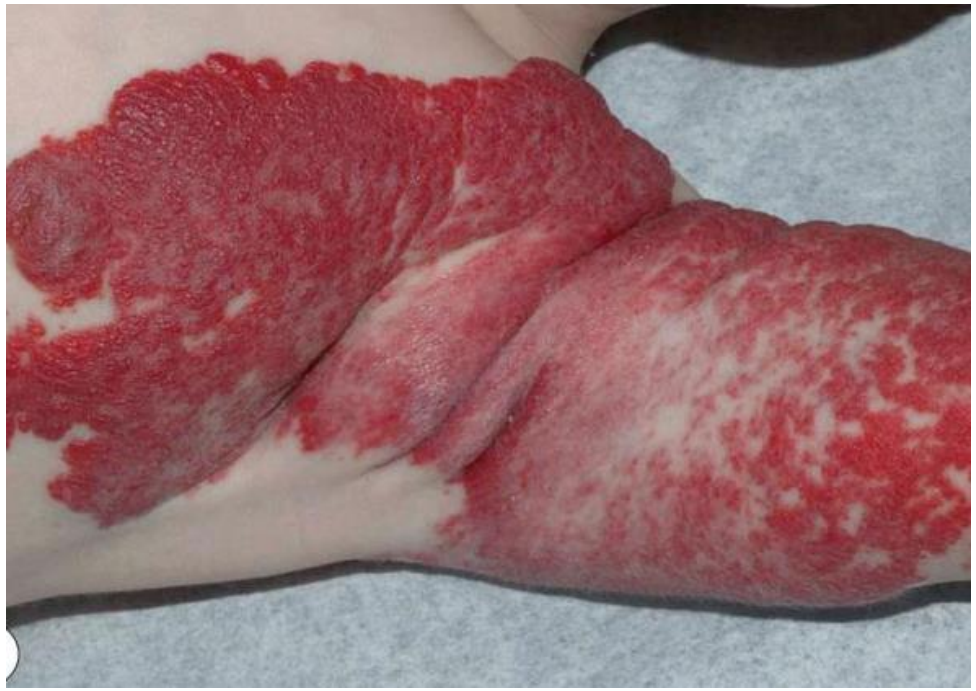




Superficial haemangioma

- **Superficial dermis**
- **Lobulated appearance**
- **Bright red**

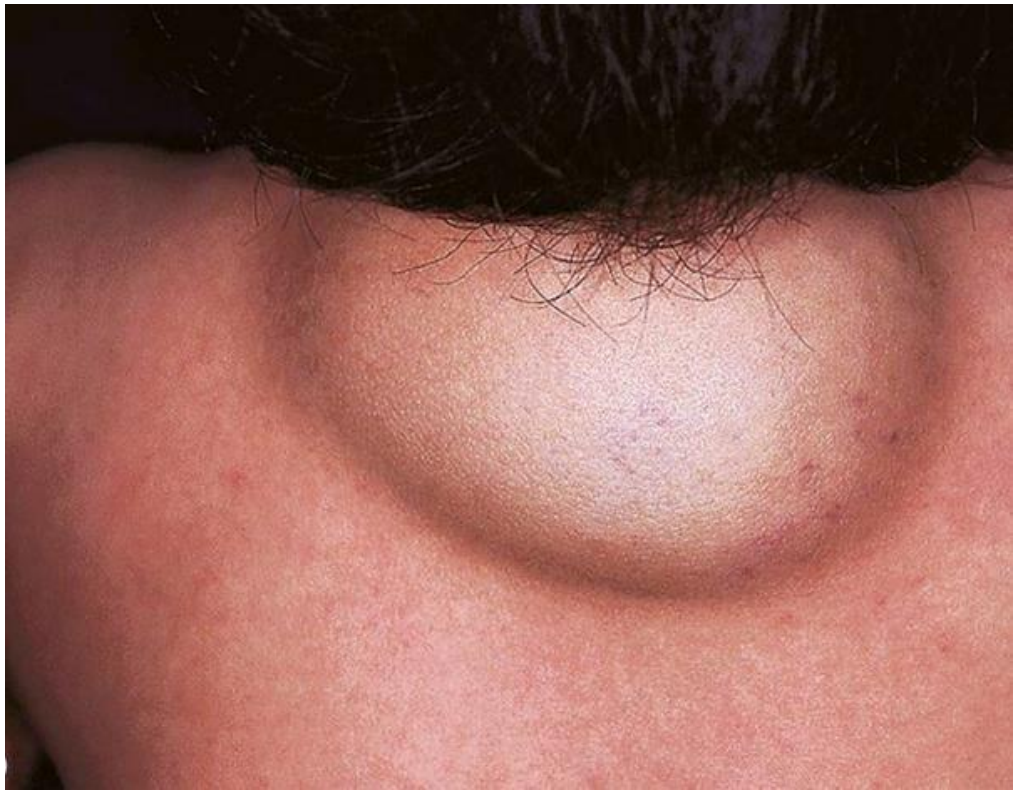


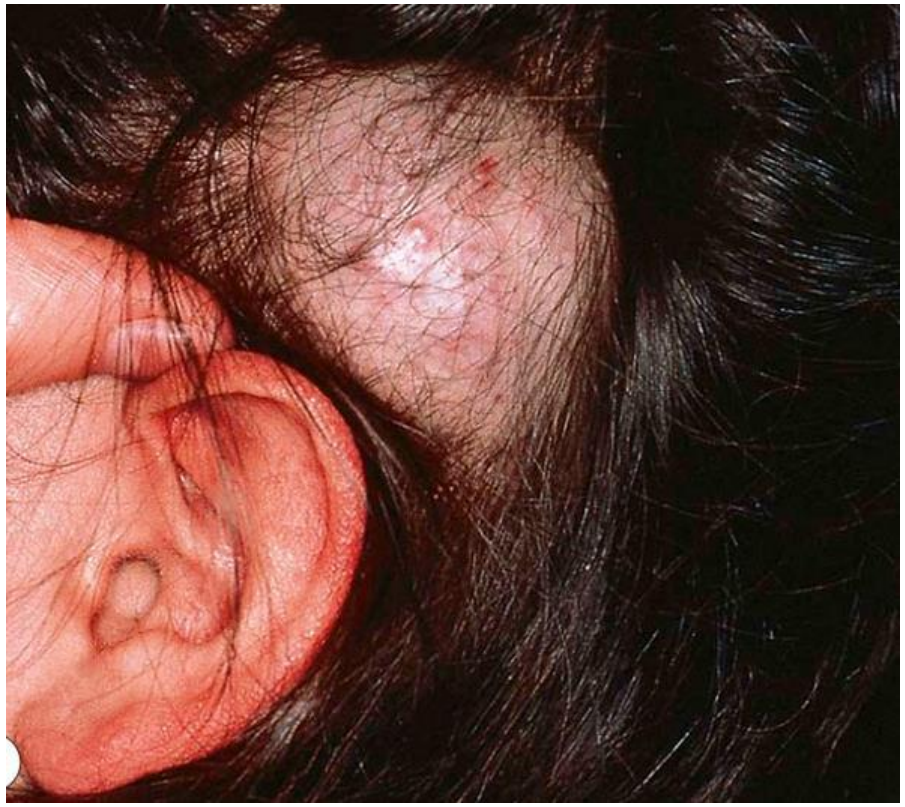


Deep haemangioma

-Deep dermis and subcutis

- Evident only after few weeks



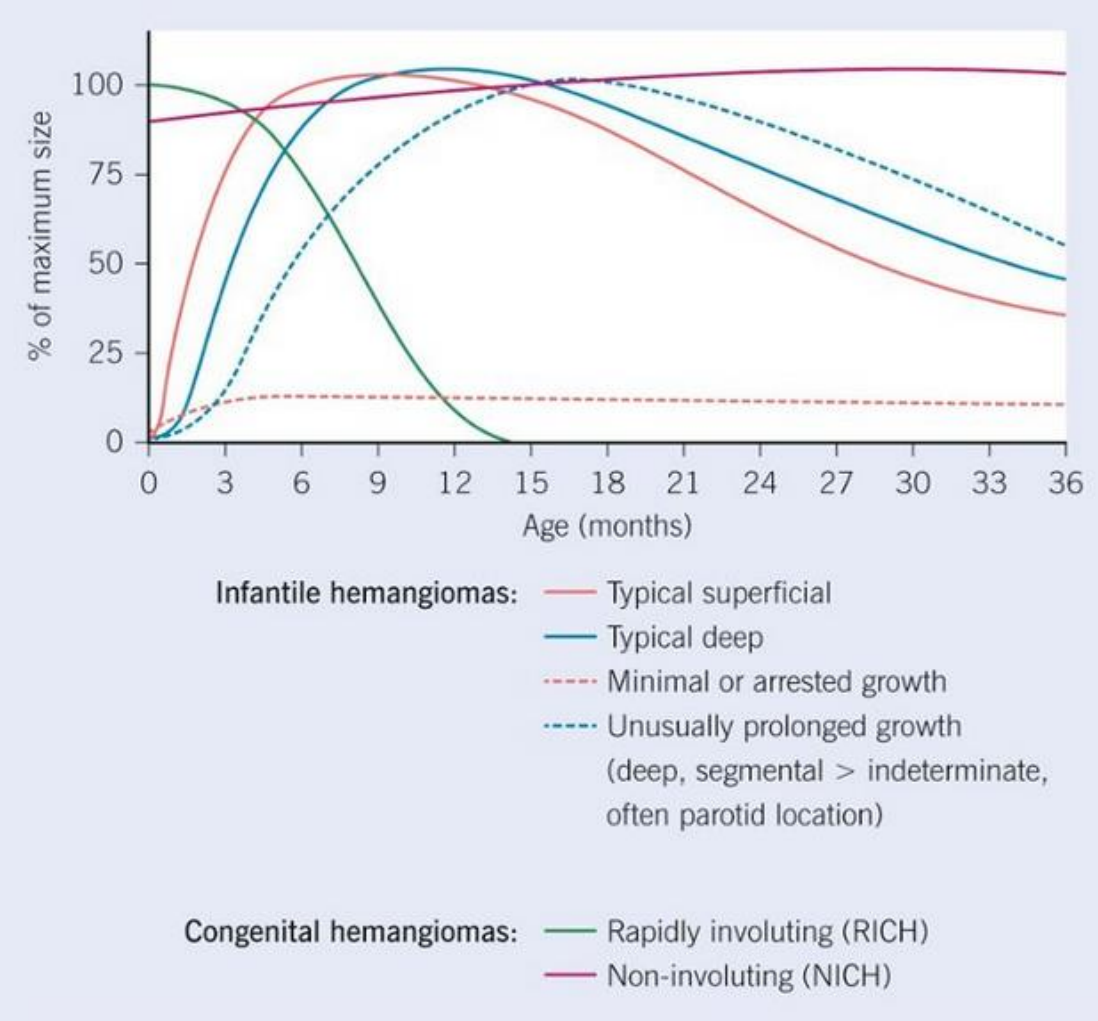


Natural history

- Early proliferative phase → Late proliferative → Plateau → Involute

- 80% of haemangiomas achieve final size by end of early proliferative phase –median 3.2 months

- Usually growth complete and involution commenced by 12 months



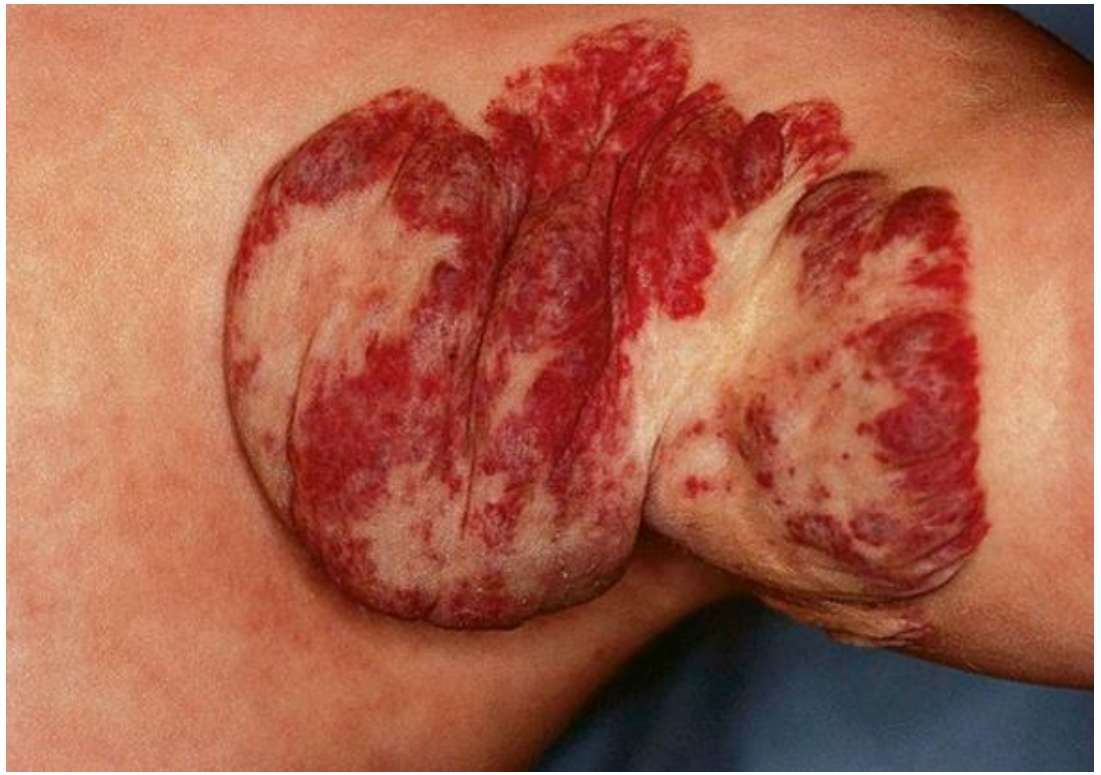
What does involution look like ?

- Change to from bright red to grey purple
- Flattening
- Less firm, more fatty

- 50% involuted by age 5 years

- 70% involuted by age 7 years

- 90% involuted by 9 years



What are the main complications ?

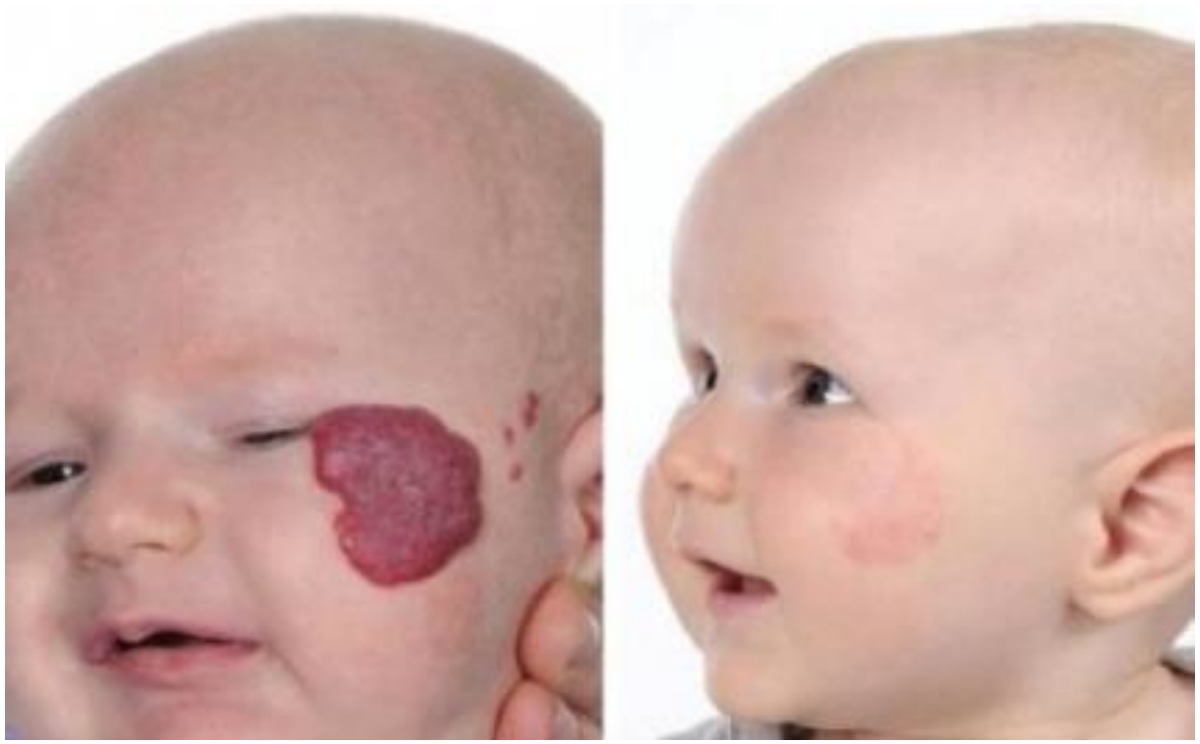
- Ulceration (10%)
  - Risk of infection and scarring
- Disfigurement and interference with function due to location or large size
  - Periocular
  - Lip
  - Nasal tip
  - Pinna
- Systemic involvement

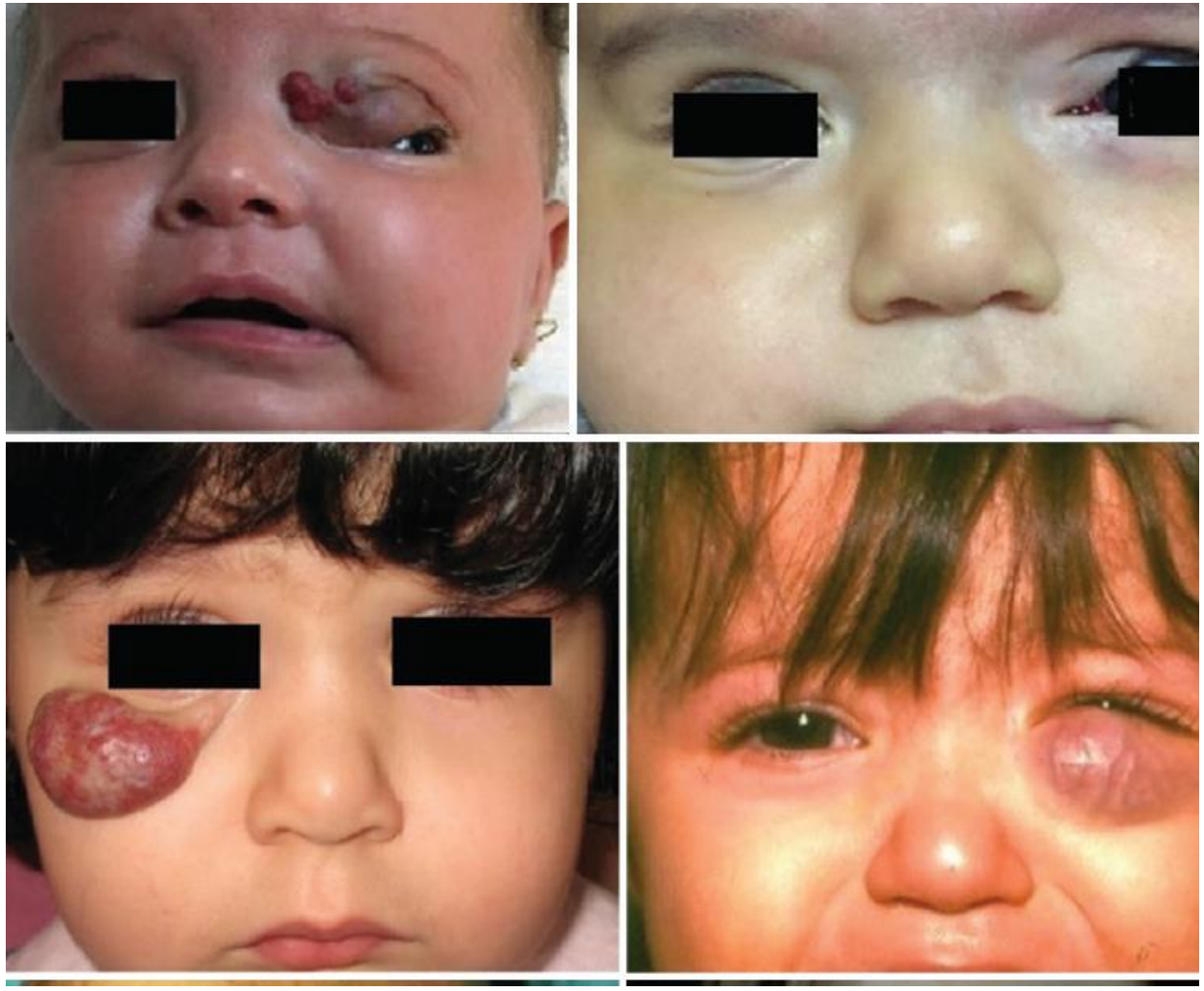
Who should be referred ?

Who should be referred ?

- Function threatening
  - Periocular
  - Nasal tip
  - Ear (extensive)
  - Lips
  - Genitalia/perineum
- Large facial
- Large anogenital/perineal
- Lumbosacral
- “Beard” distribution
- Ulcerating
- Multiple
- Diagnostic doubt

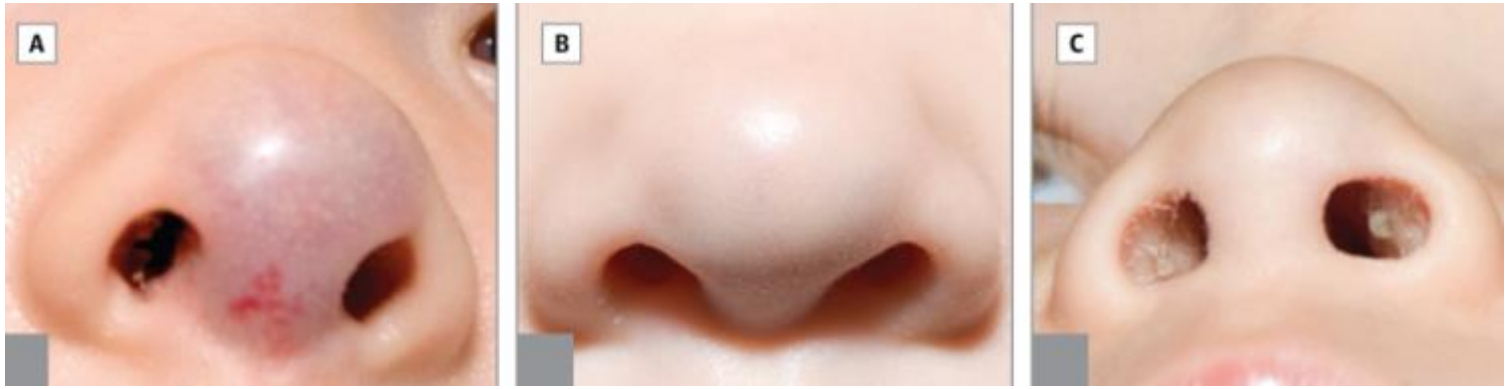
Potential for threatened vision

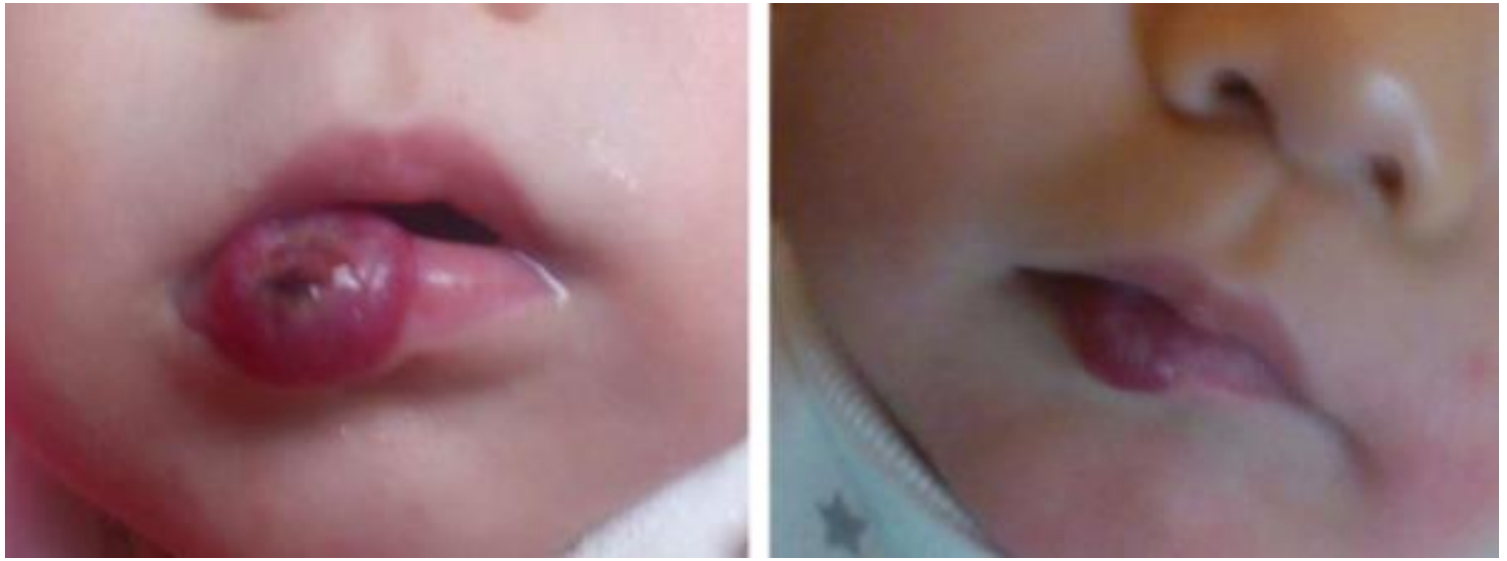




Potentially disfiguring sites

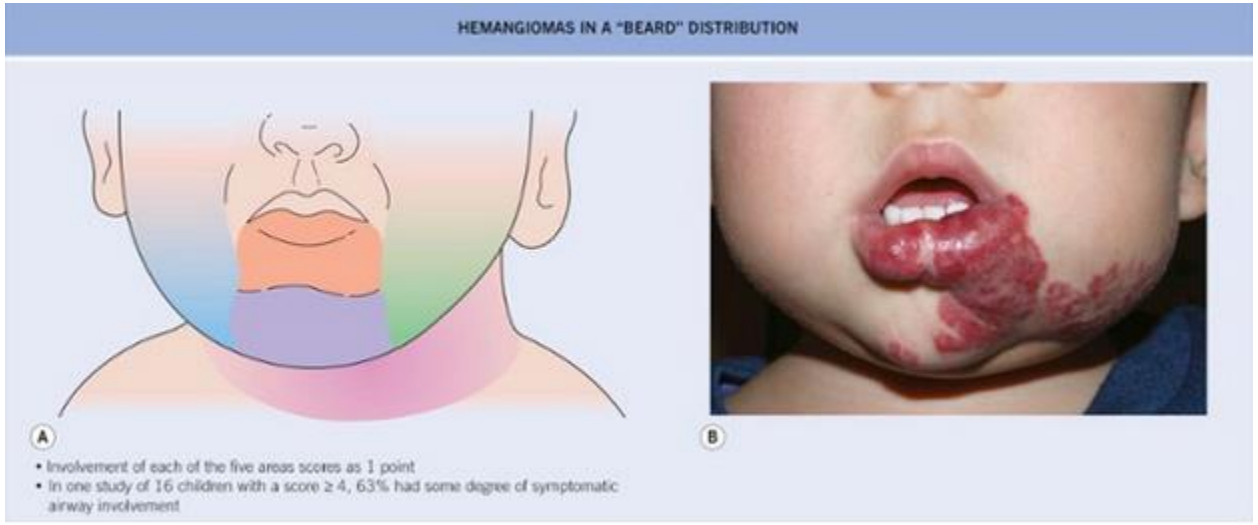
- Nasal tip
- Lip
- Rapidly expanding lesions on the face





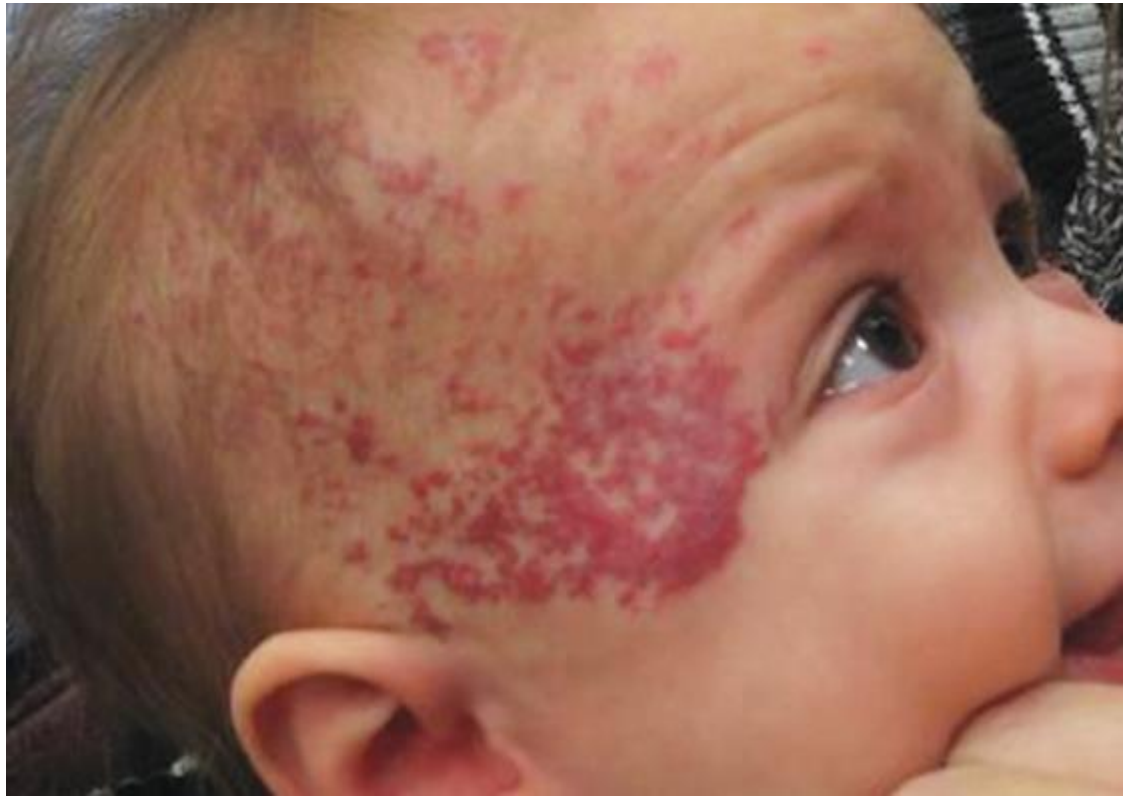
When else should I worry ?

- **<u>Lower facial or “beard” haemangiomas</u>** → marker of laryngeal haemangioma
- Risk of airway haemangioma
- URGENT assessment via ENT for assessment of airway



PHACES syndrome

- **Facial haemangioma >5cm plus…**



- **P**osterior fossa malformation
- **H**aemangioma
- **A**rterial anomalies of cervical/central

Vessels

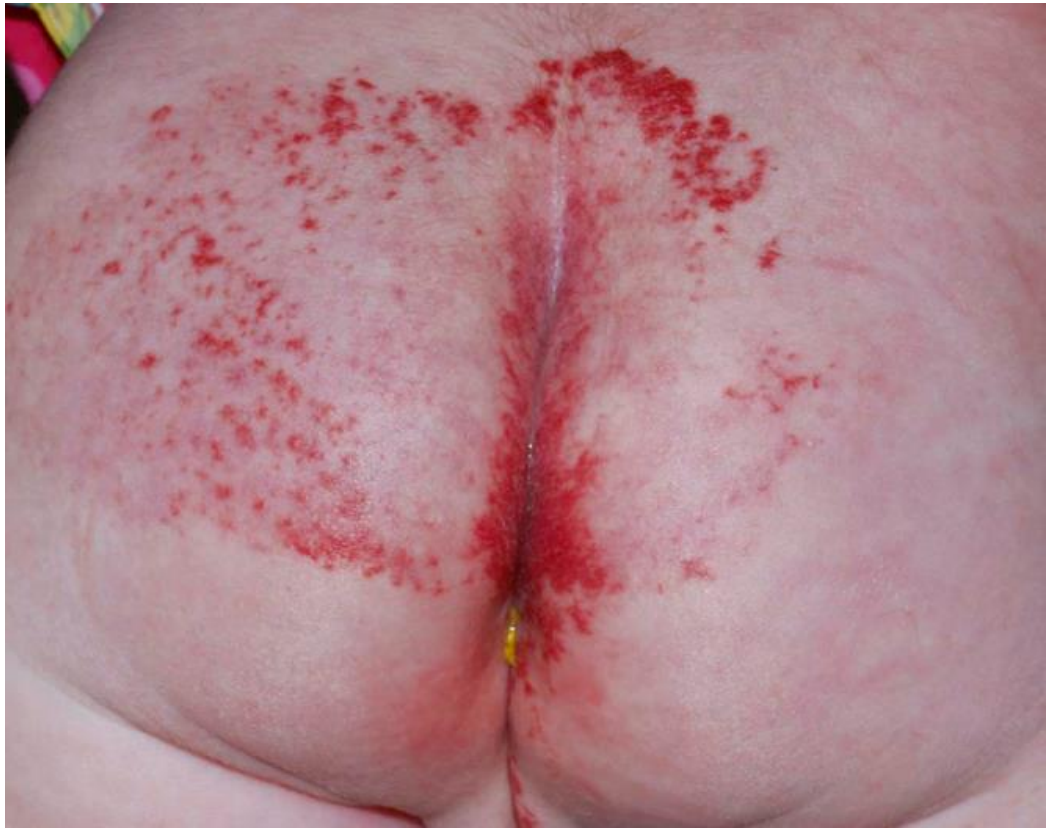
- **C**ardiac defects
- **E**ye anomalies
- **S**ternal defects and supraumbilical raphe

When else should I worry ?

- **Large segmental haemangiomas on the lower body**

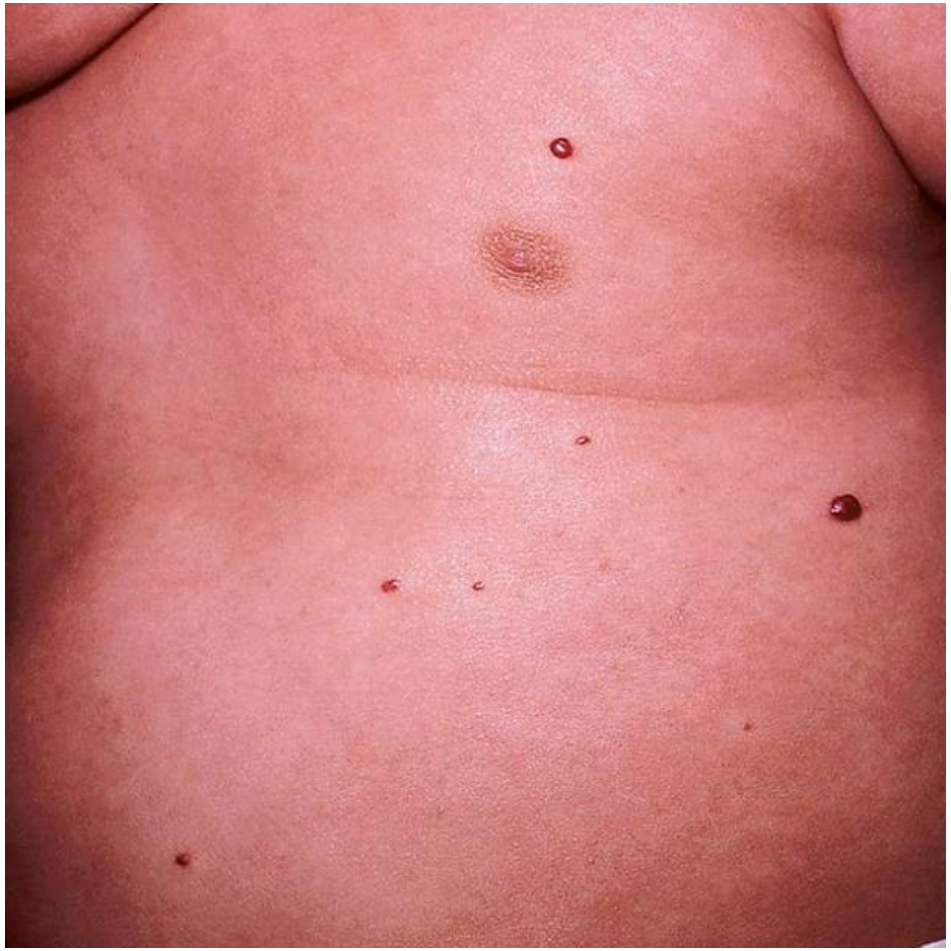
(PELVIS/SACRAL/LUMBAR)

- **L**ower body or lumbosacral haemangioma and lipomas
- **U**rogenital anomalies and ulceration
- **M**yelopathy
- **B**ony deformities
- **A**norectal and arterial anomalies
- **R**enal anomalies



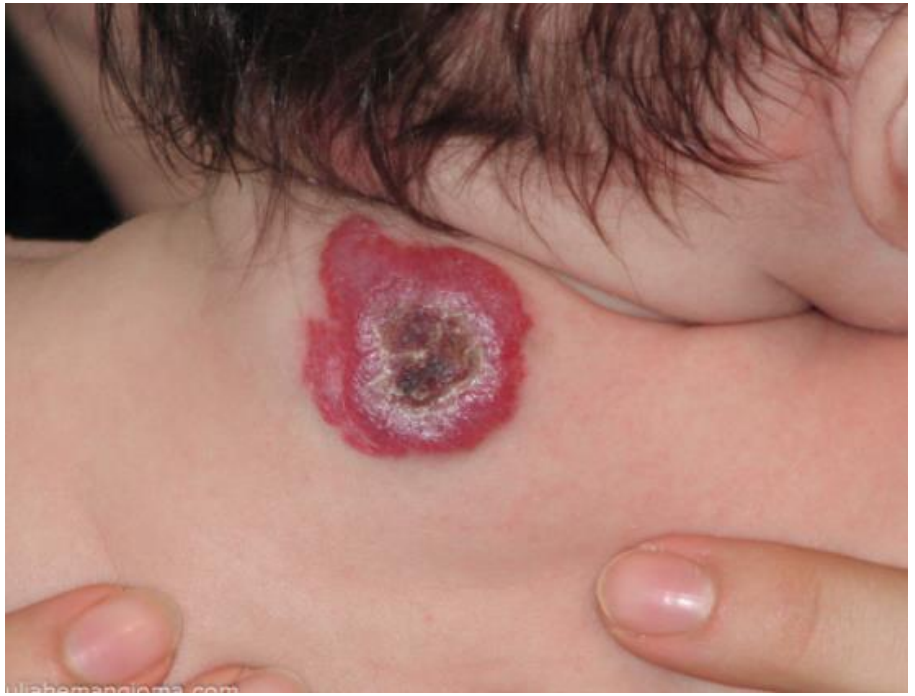
When else should I worry ?

- **<u>Multiple lesions</u>**
- Found in 10-25% of patients
- If > 5 lesions → need to investigate for visceral involvement
- Liver is most common site of involvement (hepatic haemangiomas in 16% of those with 5 or more)
- Multiple other organs can be involved
- If large/diffuse liver involvement → serious complications



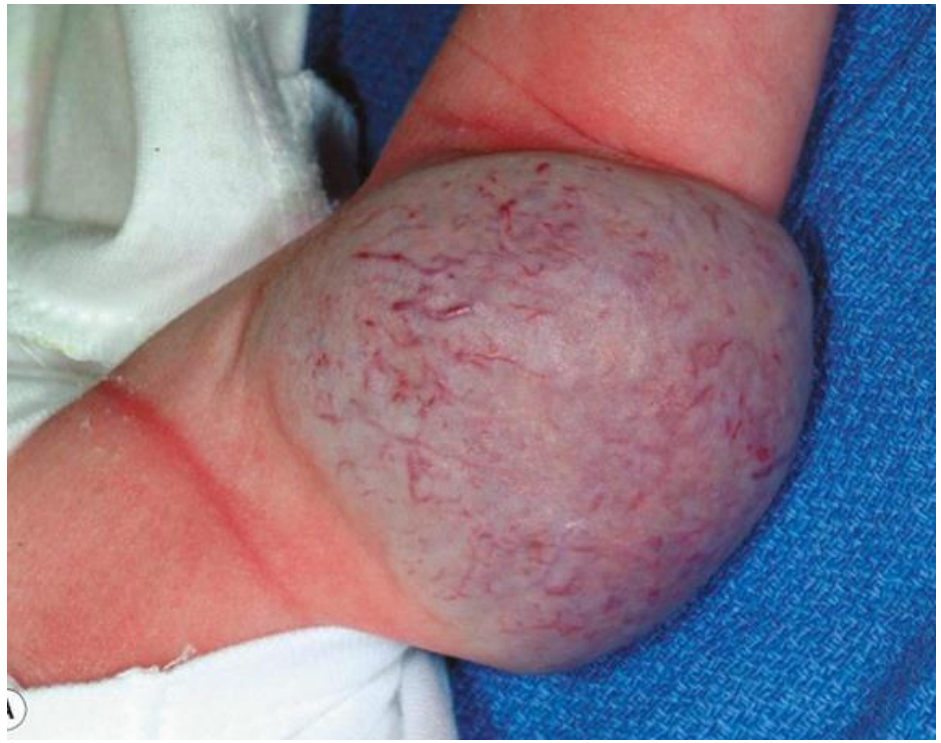
Treatments for ulcerated lesions

- Most common complication (occurs in 16% of patients)
- Most commonly occurs during proliferative phase
- Common on lips, areas of trauma
- Topical mupirocin, non-adherent wound dressings
- Oral abx if persistent/deep ulceration
- Pain relief
- May warrant systemic treatment
- GOSH guidance on washing and dressing ulcerated haemangiomas



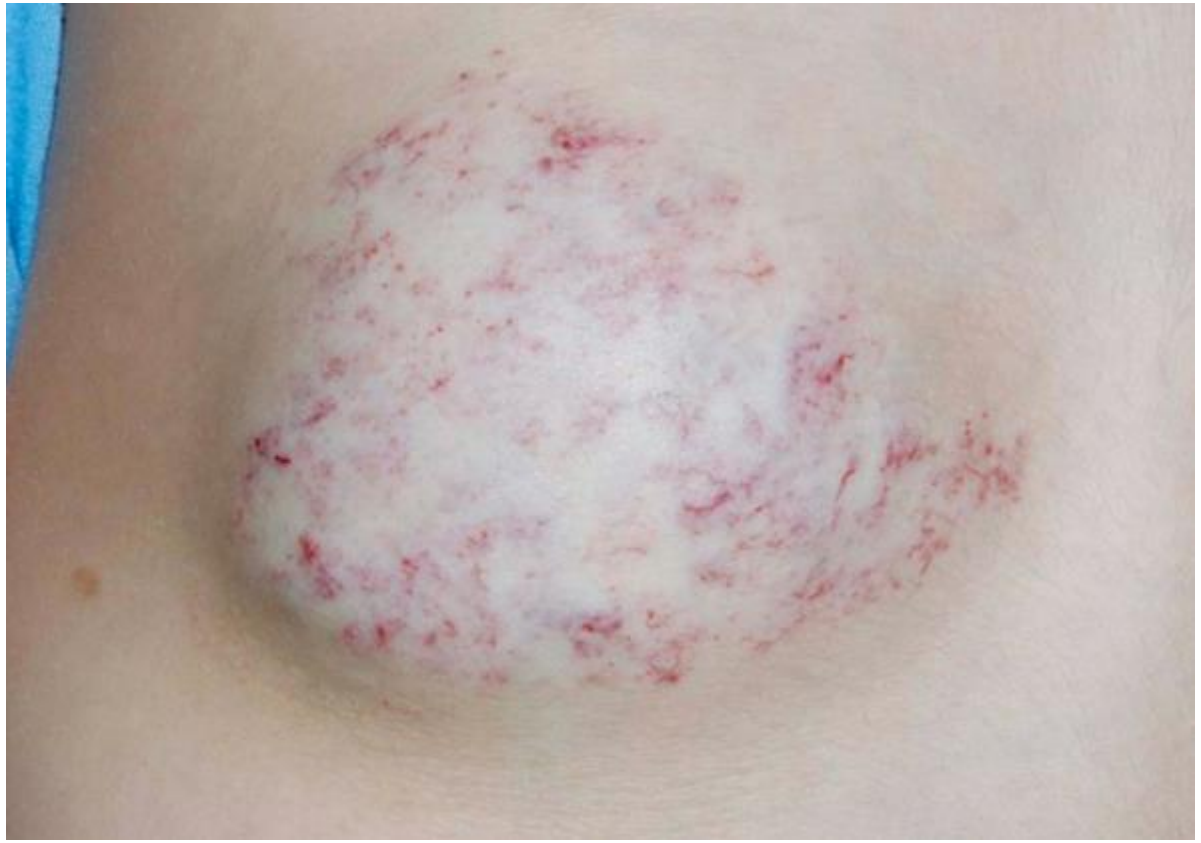
RICH vs NICH

- Rarely have a fully developed haemangioma at birth
- Clinically and pathologically distinct from IH
- Rapidly involuting capillary haemangioma (RICH)
  - Significant intrauterine growth and rapid involution by 1 year
  - Evident on USS at 12 weeks
  - Ulceration, necrosis within tumour
  - Transient ↓ platelets



RICH vs NICH

- **Non involuting capillary haemangioma**
- Lesions may be diagnosed in utero
- More often in males
- Warm to palpate and grow proportionally with the child
- Lesion may worsen with maturity



Radiology

- USS if uncertain of diagnosis of deep haemangioma, minimal overlying skin changes

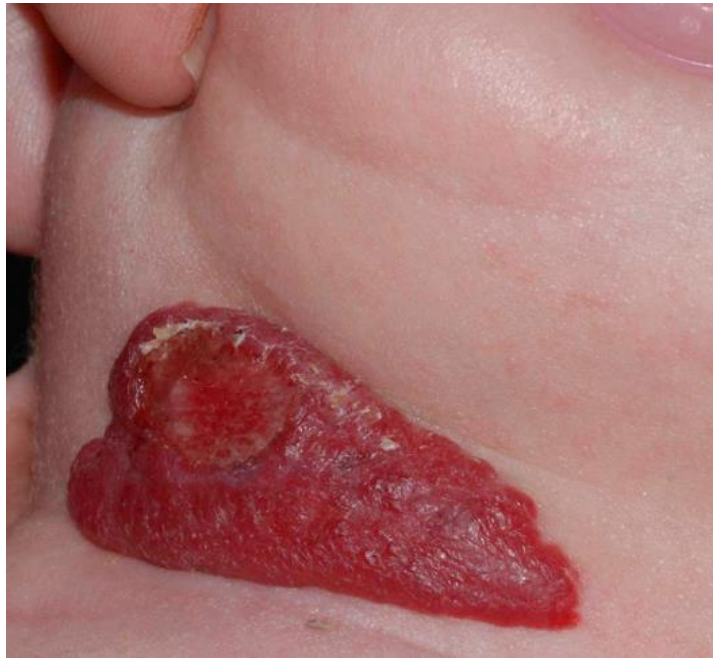
- MRI +/- MRA most useful investigation to delineate and diagnose vascular tumours

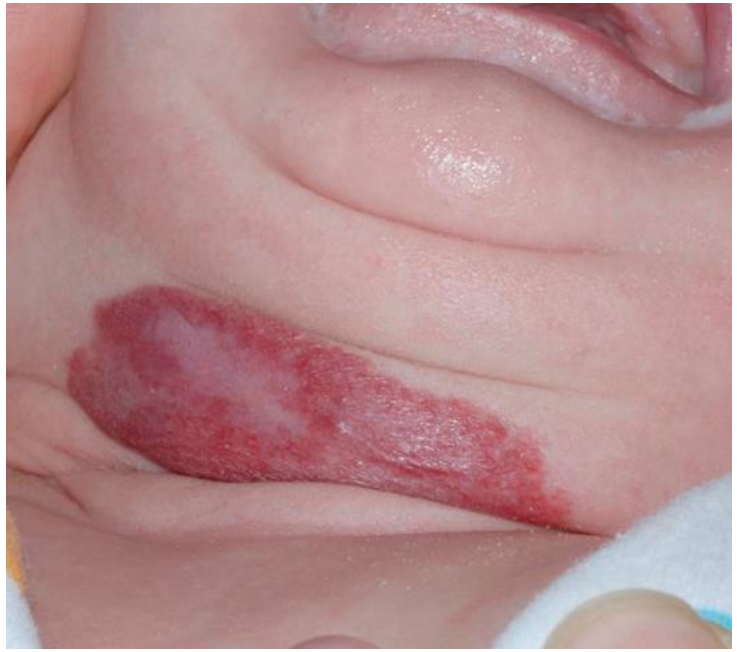
Management

- Active non-intervention
  - Photography
  - Support groups (Vascular birthmark foundation www.birthmark.org)
- Management of ulceration
  - Local wound care
  - Treatment of infection
  - Pulse dye laser
  - Surgery
  - Pain relief

Local therapies

- Intralesional steroids
- Topical steroids
- **Topical timolol 0.5% gel**





Topical Timolol

- Widely used in paediatric glaucoma
- Effective in small, superficial IH
- Beta-blocking potency = 8 x propranolol
- Wiebel L *et al* Ped Derm 2016
  - 40 infants median age 18 weeks
  - Median size 3 cm2
  - 0.5% timolol gel bd
  - 87.5% regressed, 3 continued to grow. 9 ulcerated lesions healed in 14 days
  - **Urinalysis positive** in 20/24 including all ulcerated ones
  - No significant side effects
  - Conclusion : OK for small , non ulcerated lesions – if larger may as well go for propanolol

Systemic therapies

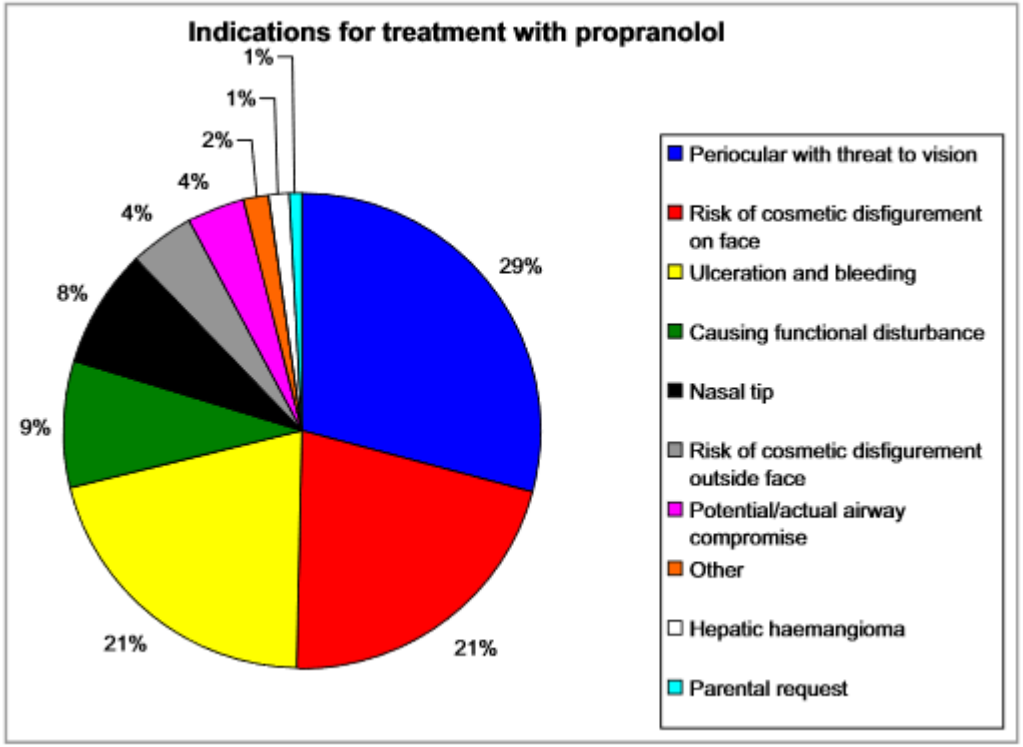
- Systemic corticosteroids
  - 2-3 mg/kg doses, need to continue whilst in proliferative phase
  - Response rate of 84%, risk of adverse effects particularly with 3mg/kg dose
  - Hypertension, HPA axis suppression, vaccination issues
- Oral propranolol
  - Original serendipitous discovery in 2008 – highly efficacious
  - More favourable side effect profile
  - Bradycardia, hypotension, hypoglycaemia, sleep disturbance, bronchospasm

Br J Dermatol. 2015 Oct 16. doi: 10.1111/bjd.14233. [Epub ahead of print]

Propranolol in the treatment of infantile haemangiomas: Lessons from the European Propranolol In the Treatment of Complicated Haemangiomas (PITCH) Taskforce Survey.

Wedgeworth E[1], Glover M[2], Irvine AD[3], Neri I[4], Baselga Torres E[5], Clayton TH[6], Beattie PE[7], Bjerre JV[8], Burrows NP[9], Foelster-Holst R[10], Hedelund L[11], Hernandez-Martin A[12], Audrain H[13], Bhate K[14], Brown SJ[15], Baryschpolec S[16], Darne S[17], Durack A[9], Dvorakova V[3], Gach J[18], Goldstraw N[19], Goodyear H[20], Grabczynska S[21], Greenblatt D[19], Halpern J[22], Hearn RM[15], Hoey S[23], Hughes B[16], Jayaraj R[24], Johansson EK[25], Lam M[14], Leech S[26], O'Regan GM[3], Morrison D[27], Porter W[28], Ramesh R[22], Schill T[29], Shaw L[13], Taylor AE[26], Taylor R[30], Thomson J[1], Tiffin P[31], Tsakok M[32], Janmohamed SR[33], Laguda B[34], McPherson T[32], Oranje A[35], Patrizi A[4], Ravenscroft J[14], Shahidullah H[36], Soloman L[2], Svensson A[37], Wahlgren CF[38], Hoeger PH[29], Flohr C[1].

- Data from 8 European countries, 1096 children
- Main indicators for treatment were periocular, risk of cosmetic disfigurement and ulceration and bleeding
- Most on 2mg/kg/day
- 91% had good or excellent response to Rx
- Rebound growth occurred in 14% on stopping



- 19.6% of patients experienced side effects, only 20% of whom necessitated stopping treatment
- AE noted at higher dose >2mg/kg
- No difference in adverse effects those screened with echo/ecg

Adverse event	Patients among total cohort, n (%)
Sleep disturbance	90 (8·2)
Cold peripheries	51 (4·7)
Wheezing	31 (2·8)
Diarrhoea	21 (1·9)
Symptomatic hypotension	18 (1·6)
Symptomatic hypoglycaemia	8 (0·7)
Symptomatic bradycardia	6 (0·5)
Other	36 (3·3)

The use of propranolol at SGH

- Trust guidelines with colleagues in General Paediatrics and ENT
- Jungle ward for 4 hour observation of first dose
- Subsequent monthly – three monthly reviews in OPD
- No need for community BP monitoring

- Only needed for indications described, most IH can be managed with active non-intervention

Thank you