



# Successful management of collodion baby

***Presenter: Dr. Deepika***

*Senior Resident (DM Neonatology)*

*AIIMS*

*New Delhi*

# Moderators

Dr. Ankit Verma

Assistant Professor

Department of Pediatrics

AIIMS

New Delhi

Dr. G. Sethuraman

Professor

Department of Dermatology

AIIMS

New Delhi

# Baby P

- 34 weeks, 2.6 kg, male baby
- Born on 25.09.2019
- **Obstetric history:** G<sub>3</sub> P<sub>0</sub> L<sub>0</sub> A<sub>2</sub>
- **Antenatal period:** Regular ANC visits, Level II USG: Normal
- Spontaneous onset of labor and leaking at 8.5 months POG
- LSCS i/v/o transverse lie (outside), cried immediately after birth

**Baby presented with.....**

# Baby P

**Severe ectropion  
and eclabium**

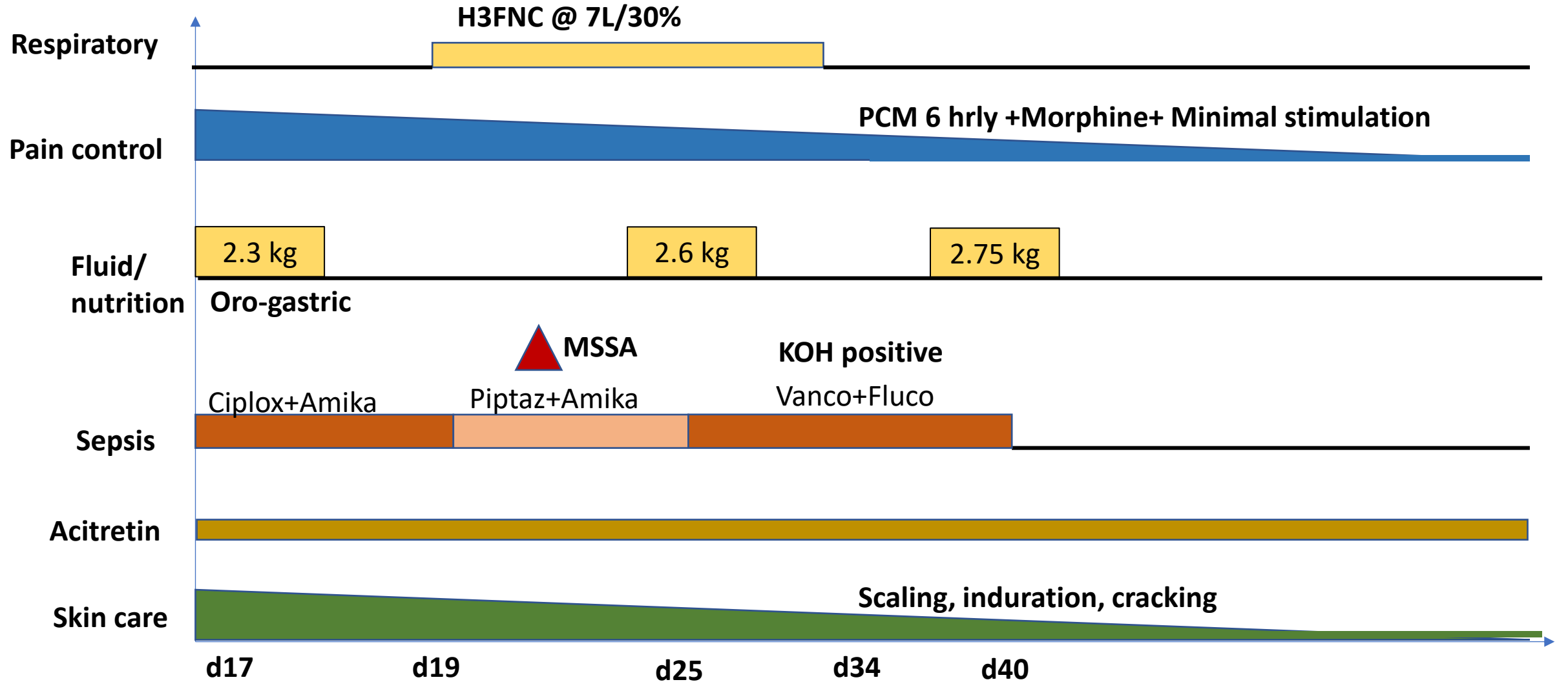


**Thick membranous scaling  
with areas of denudation and  
ulceration**

**Diagnosis:  
Collodion baby**

- No consanguinity
- H/o ichthyosis in 2<sup>nd</sup> cousin
- Taken to private practitioner
- Prescribed topical antibiotic and steroids
- Formula feeding via paladai at home
- Presented to AIIMS on day 17

# Baby P



# Day 42 of life

- New onset fever
- Limitation of right lower limb
- Marked tenderness
- Striking asymmetrical swelling



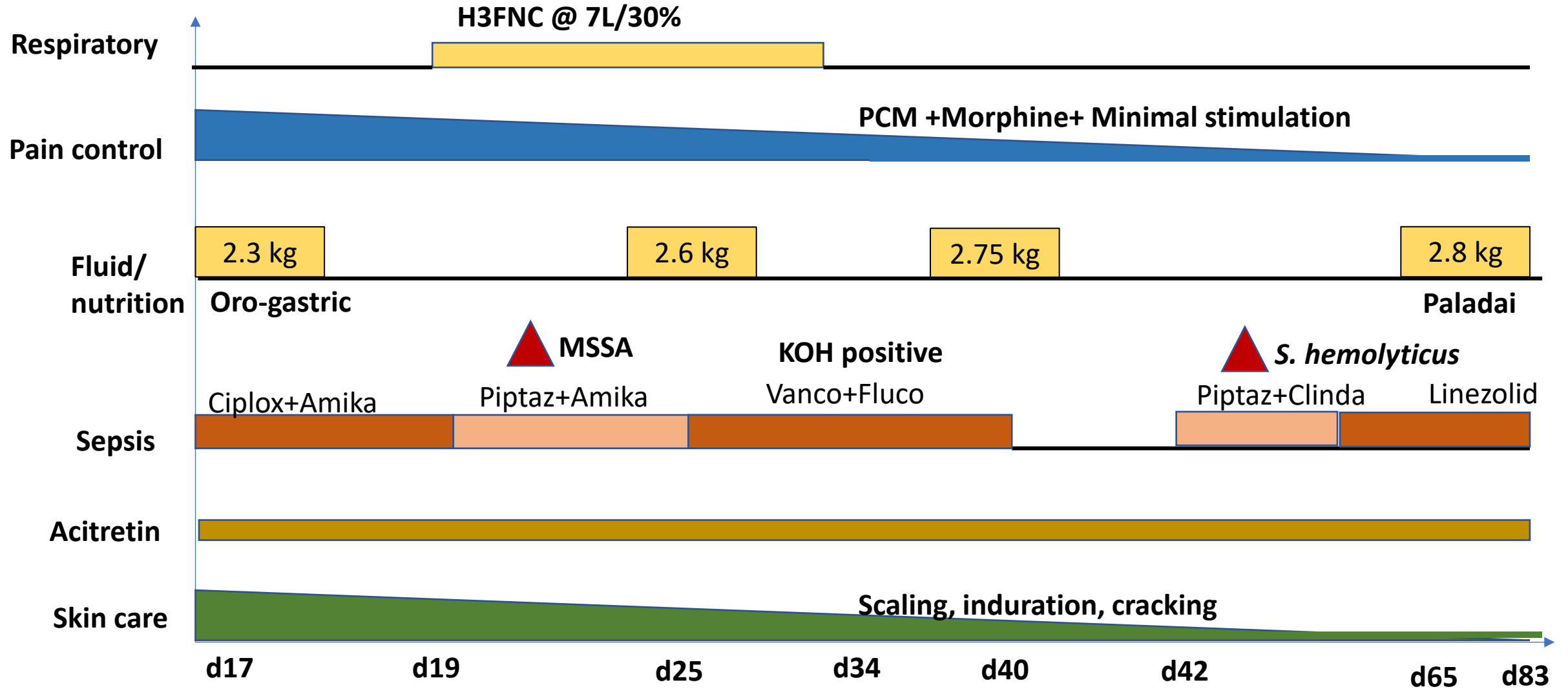
**Possibilities??**



# Day 42 of life

Possibilities	Points in favor	Points against	Action
1. Osteomyelitis	Two episodes fever+ Asymmetric Swelling	Unusual in neonates	X Ray ( <b>normal</b> ) USG hip ( <b>No soft tissue swelling/erosion/widening</b> )
2. Septic arthritis	Decreased mobility Blood c/s positive MSSA		Sepsis screen ( <b>Positive</b> )
3. Painful skin induration	Dynamic contractures + Healing phase of collodion baby	Asymmetric (R>>L)	Already on analgesics New onset
4. Trauma	Unilateral	No h/o evident trauma	-

# Baby P





# Genetic counselling

- Advised for genetic testing as **“Targeted sequencing”** for implicated genes
- Outsourcing of test and non-affordability of parents
- Advised antenatal genetic testing and counseling in the next pregnancy



**Birth**



**Day 50**



**At discharge**



**10 months**

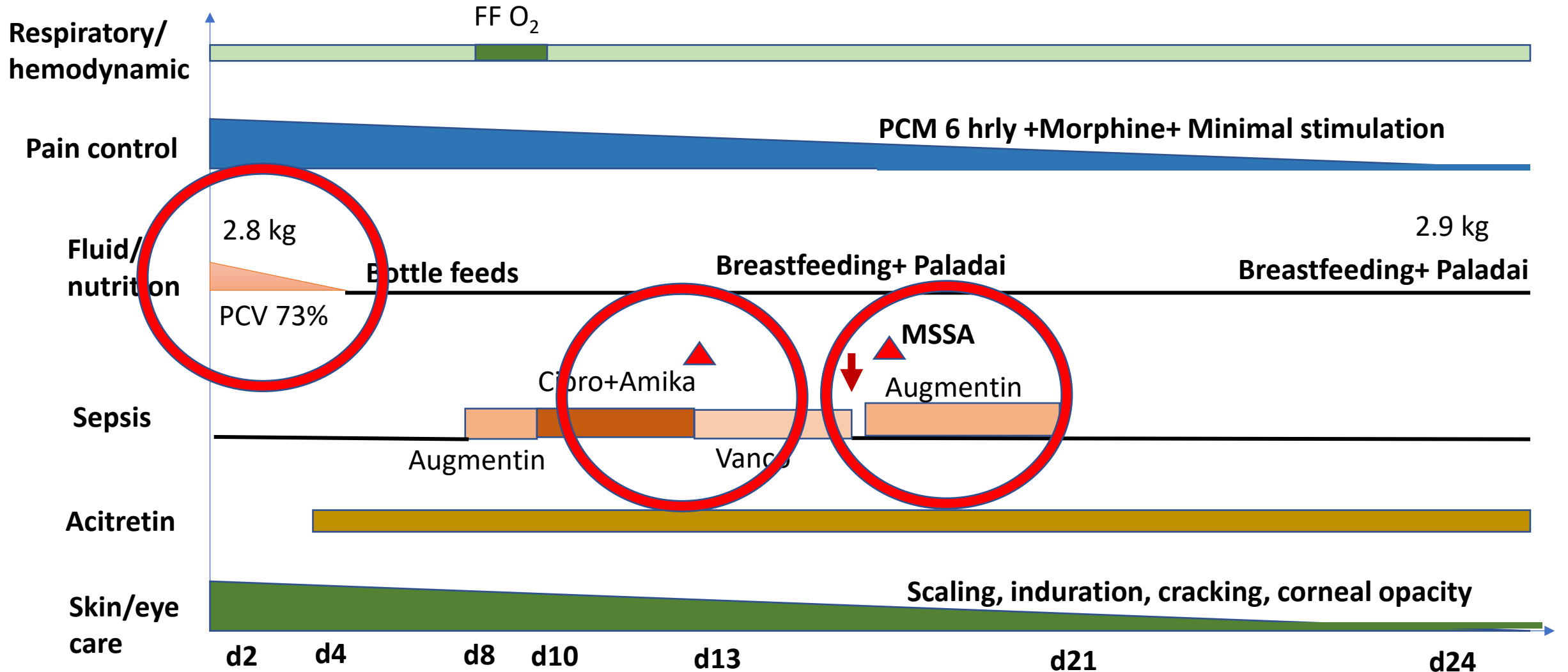
# Baby R

- Term, 2.8 kg, male baby
- Born on 26.11.2019 vaginally
- Cried immediately
- **Obstetric history:** Primigravida
- **Antenatal period:** Uneventful
- Level II USG: Normal
- Admitted to AIIMS at d2 of life
- Third degree consanguinity
- No family history





# Baby R





**Birth**



**At discharge**

# Collodion baby

- 300 cases till now
- Severe congenital ichthyosis
- '*Ichthys*' means fish
- Mostly autosomal recessive
- Taut, shiny, cellophane-like membrane, ectropion and eclabium

**'Dipped in hot wax'**





# Pathogenesis

Genetic mutation

*TGM1 (40%),  
ALOX12B, ALOXE3*

Abnormal and excessive  
Cornification

*“Deficient cross-linking of  
cornified cell envelope proteins”*

Disruption of skin barrier

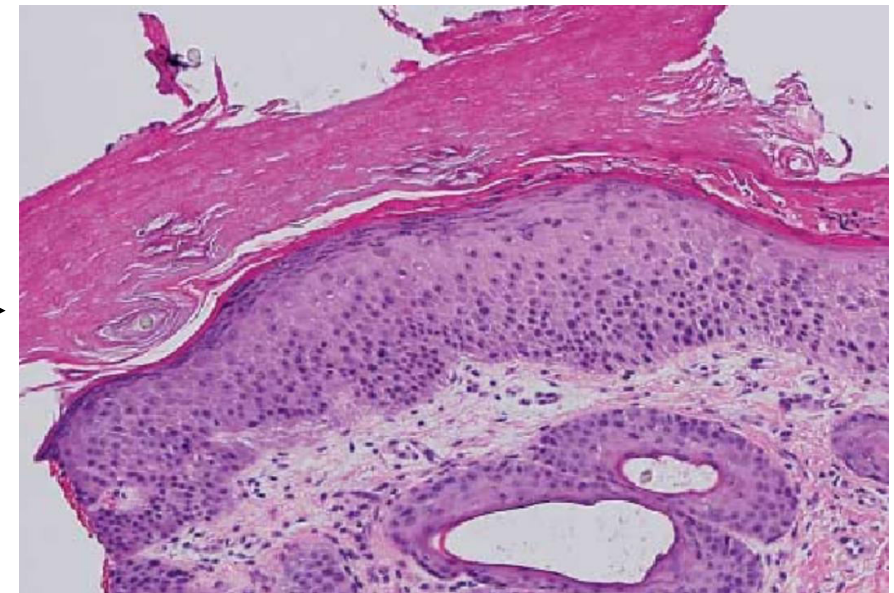
*Sepsis*

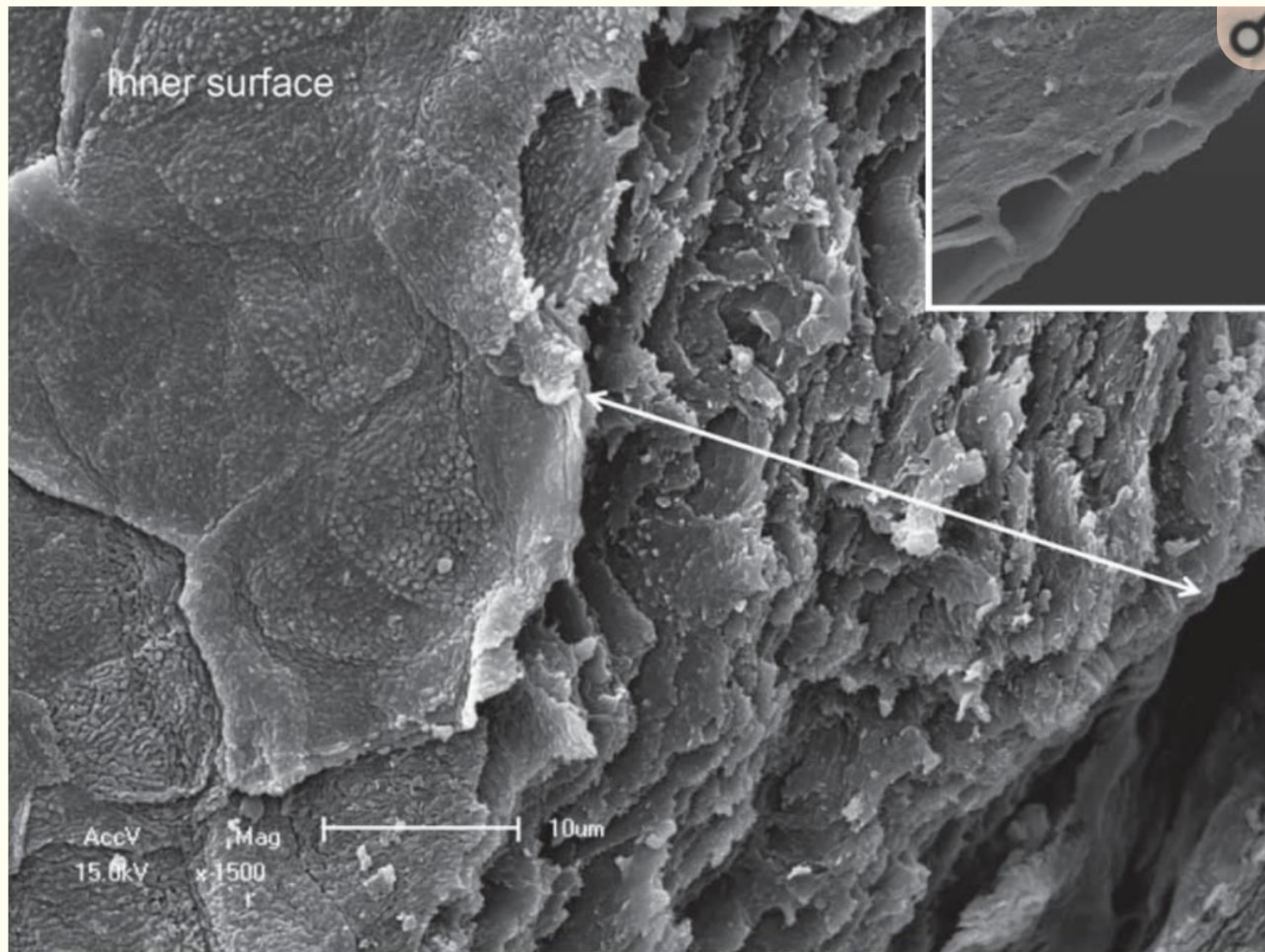
*Temperature instability*

*Increased TEWL*

*Hypernatremia*

normal





**FIGURE 3**

Scanning electron microscopy - lateral view of the collodion membrane showing multilayered corneocytes (x1.500). Inset with normal skin showing the “basket weave” aspect of the horny layer (x1.200).



# Collodion baby: Outcome

**“Phenotypic change, membrane usually sheds within 3-4 weeks”**

## Non-syndromic

1. Lamellar ichthyosis (LI) (50%)
2. Non bullous congenital ichthyosiform erythroderma (NBCIE) (20%)
3. X-linked ichthyosis
4. Ichthyosis vulgaris

## Syndromic

1. Lipid storage disorder
2. ARC syndrome
3. Others

## Metabolic

1. Holocarboxylase synthetase deficiency
1. Gaucher disease type 2
2. Congenital hypothyroidism

**Self-healing collodion baby (10-25%)**

# Differential diagnosis

**Disease**  
Harlequin ichthyosis

Ichthyosis prematurity syndrome

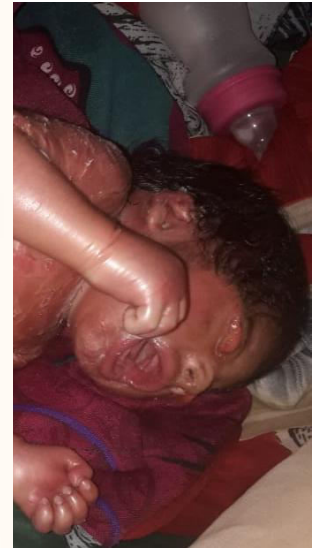
Netherton syndrome

Sjögren-Larsson syndrome

## Harlequin ichthyosis



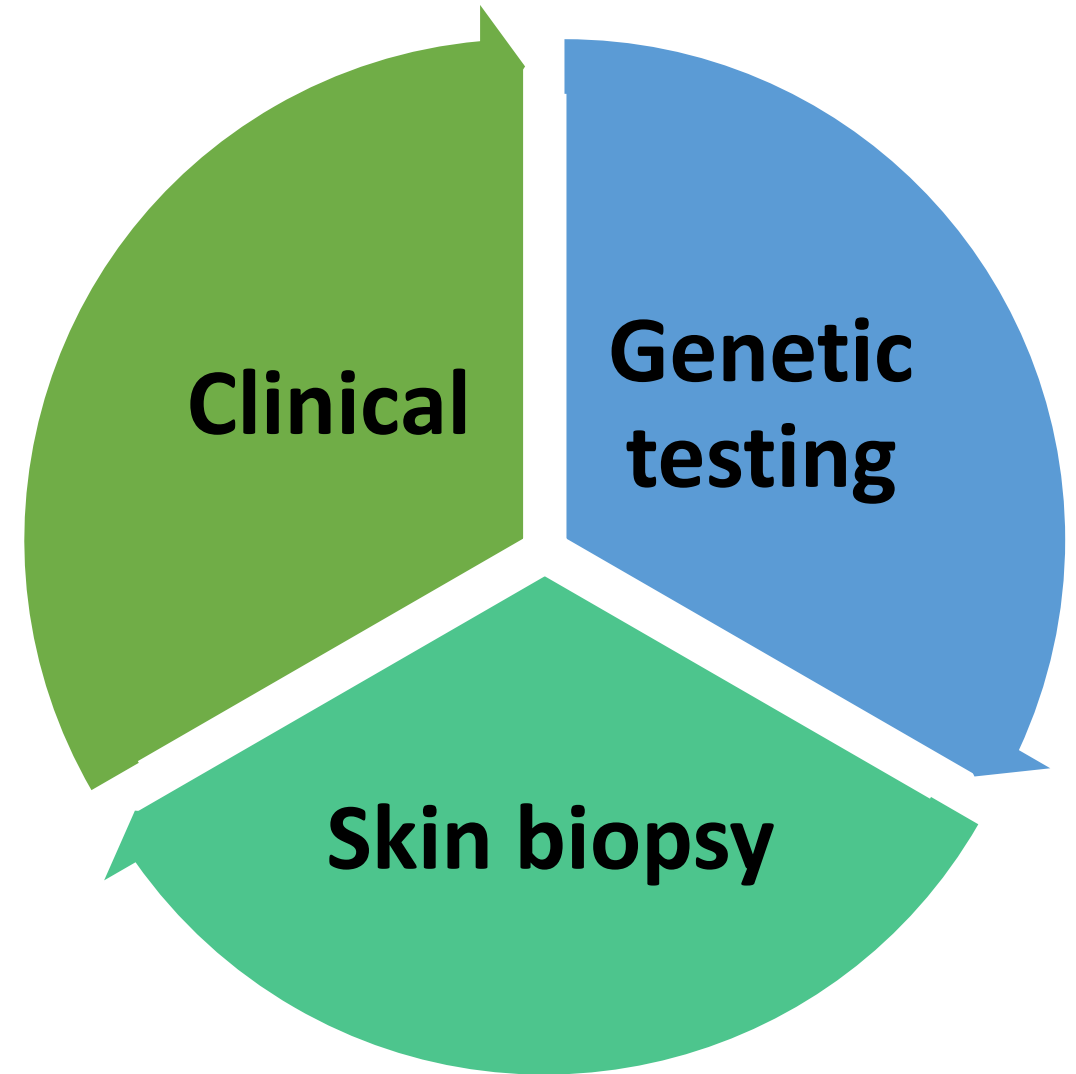
“Thick, white, armor-like scales  
Deep fissures in diamond pattern”



“like membrane”

# Diagnosis

- **Purely clinical** in cases of non syndromic ARCI
  - Typical findings at birth
- **Skin biopsy**: Not necessary
- **Genetic testing**: Multigene panel testing **12 genes**
- **Single gene testing**: ABCA12 (Harlequin), TGM1 (ARCI)



# Collodion baby: Management



**“Multi-  
disciplinary  
approach”**

# Skin care

- **Soft bedding**
- **Use of emollients:** Use of inert emollients in neonates  
Paraffin dressings and petrolatum jelly  
Too much emollients (risk of infections)
- **Topical antibiotics:** Areas of denudation, fissuring and ulceration





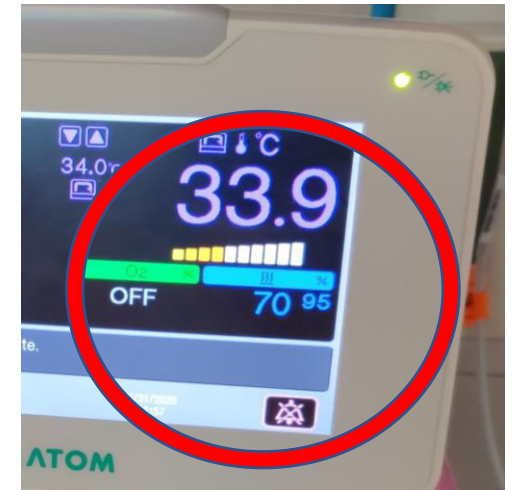
# Skin care

- **Humidified incubators:**

No standard guidelines for level and duration, 60-70% adequate

Tapered to 20% over days to weeks

- Daily sponging/bathing



# Eye care

- **Topical methylcellulose:** To prevent exposure keratitis
- **Antibiotics:** Prophylactic and therapeutic as risk of conjunctivitis very high



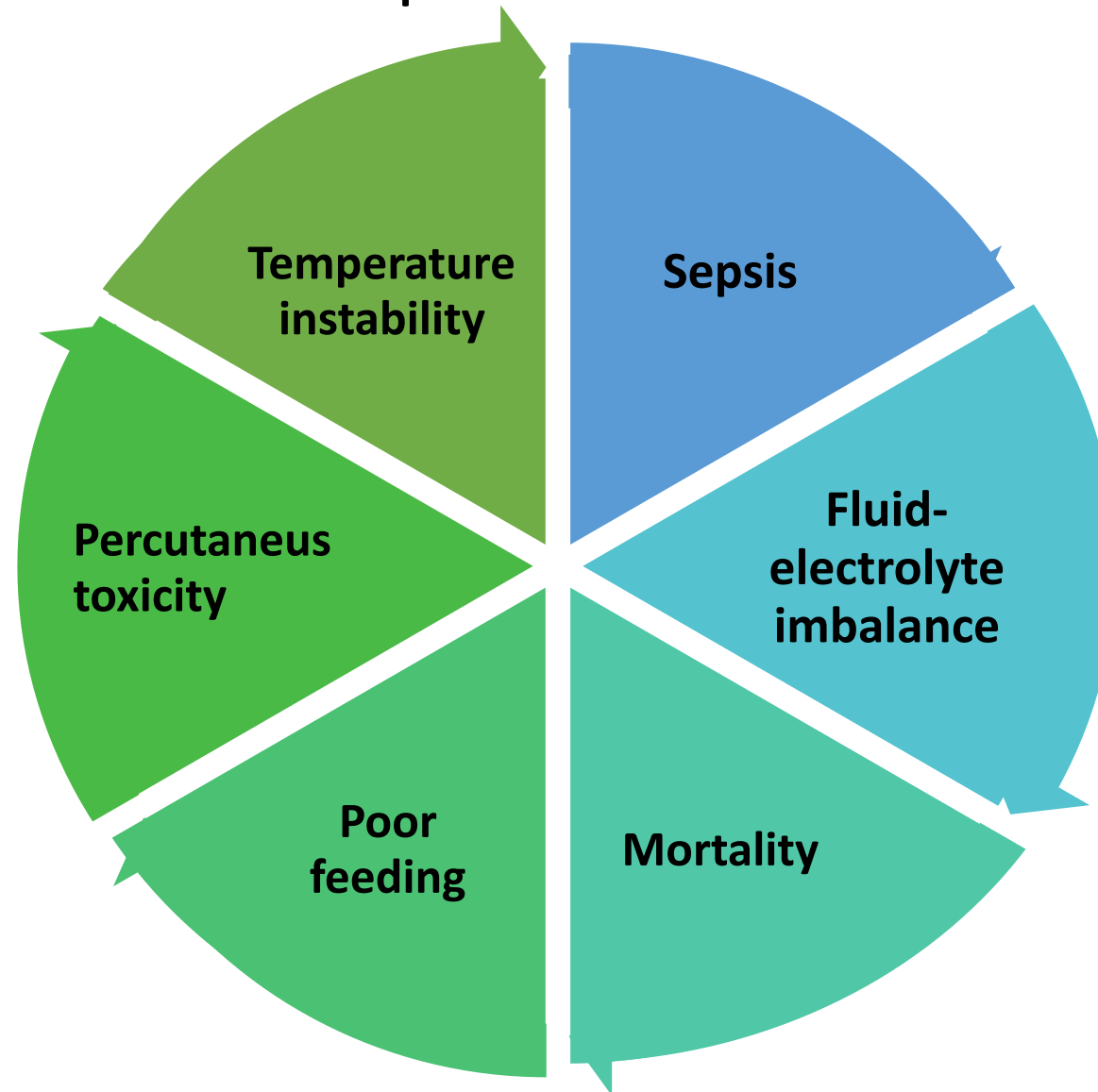
# Feeding

- Oral feeding to be encouraged
- **Orogastric feeding** if severe stiffness and pain

# Pain control

- Analgesia (Acetaminophen, opioids) with pain scoring

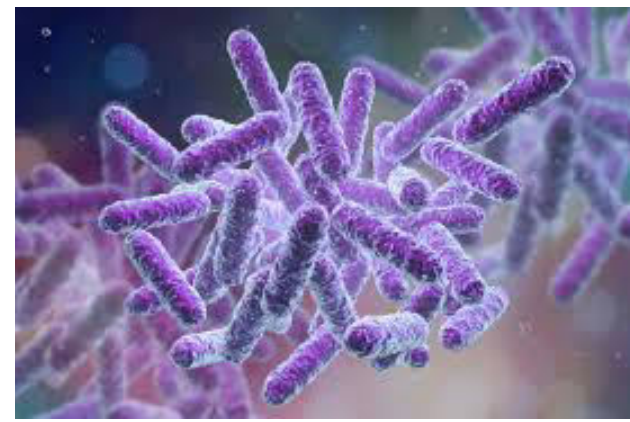
# Complications



- 1960: 50%
- 1986: 11%
- 2012: 5%



# Complications: Sepsis



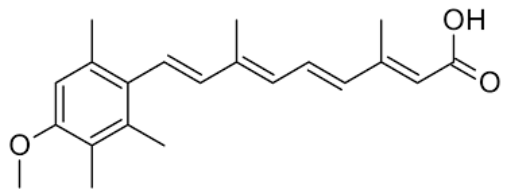
- Increased risk due to disrupted natural skin defenses
- Bacterial: *S. aureus*, *S. epidermidis*, *S. hemolyticus*, *E.coli*
- Fungal: *Candida*
- Increased propensity to bone and joint infections
- High index of suspicion, early recognition and prompt treatment
- **No role of prophylactic antibiotics**

*Van Gysel D, Lijnen RLP, Moekti SS, de Laat PCJ, Oranje AP. Collodion baby: a follow-up study of 17 cases. J Eur Acad Dermatol Venereol 2002*

# Complications

No.	Sex	Pregnancy term(weeks)	Birthweight (g)	Ectropion/eclabium	Time of desquamation (days)	Skin treatment	Complications	Diagnosis
1	M	38-40	2690	+/+	10	Ureum 10% euc.	SI	NEARLI
2	F	37	2530	+/-	12	None	-	S-L
3	F	40	3620	-/-	20	petr/lan, ur 5% cmc	SI; CO; HT	NEARLI
4	F						SI; HT	NL
5	F						SI	EARLI
6*	F						SI; HD	EARLI
7*	M						SI; HT; HD	EARLI
8	M						S; SI	EARLI
9†	M						HD; SI; CO	EARLI
10	M						SI	NEARLI
11	F						S	NL
12	M	40	3340	-/-	12	none	HT; HD	EH
13	M	35	1500	-/-	7	none	HT	NL
14	M	36	2870	-/-	NK	petr/lan	-	NL
15†	M	36	3700	-/+	15	none	-	EARLI
16	F	40	4385	-/-	21	none	-	EARLI
17	M	36	1935	+/+	-	petr/lan	-	GaucherII

**Increased TEWL (upto 8-10 times)**  
**Hypernatremic dehydration (23% cases)**  
**Hypothermia (30% cases)**  
**Local (52%) and systemic (18%)**



# Acitretin: The “wonder” drug



- 2<sup>nd</sup> gen aromatic retinoid
- Vitamin A Derivative with longer half-life
- It affects (i) cell growth and differentiation (ii) alter cellular adhesiveness (iii) exerts immunomodulatory action
- **Dose:** 0.5 to 0.75 mg/kg/d, max (2mg/kg/d), Slow tapering to 0.25 mg/kg/d and withdrawn
- Duration of therapy: 3 to 6 months
- **Transient:** Cheilitis, dry mucosae, hair loss, and fissuring of palms and soles.
- **Long term:** Skeletal, pseudotumor cerebri, liver toxicity, thyroid disorder

*Prado R, Ellis LZ, Gamble R, Funk T, Arbuckle HA, Bruckner AL. Collodion baby: an update with a focus on practical management. J Am Acad Dermatol 2012*



# Family counseling



**Hopeful  
improvement**

# Challenges faced

- Severe form with severe ectropion
- Difficult feeding, painful lesions, difficult cannulation and handling
- Culture positive soft tissue and bone-joint infections
- Failure to perform genetic analysis due to non-availability of consent

# Lessons learnt

- Collodion baby is rare and true dermatological emergency
- Life threatening in early phase with increased risk of complications
- All complications preventable by meticulous multi-disciplinary care
- Early use of acitretin proves highly beneficial
- Appropriate genetic counselling must for all cases



**Thank you**