AN INTERESTING BULLOUS DISEASE OF CHILDHOOD

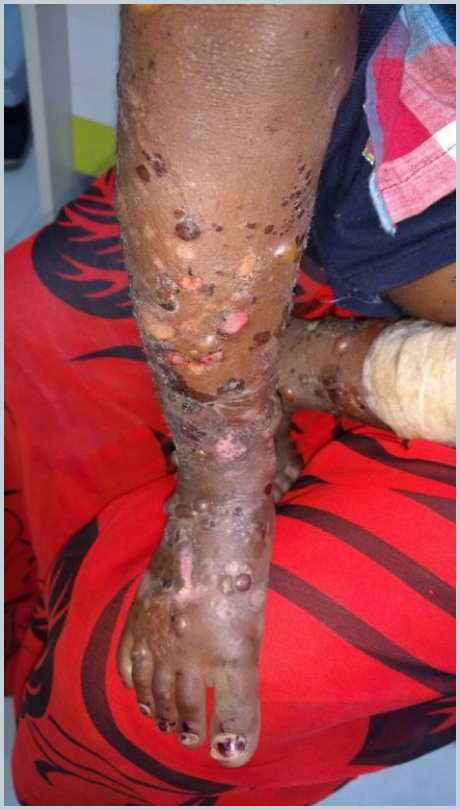
DR.REVATHY.S, REGISTRAR IN PEDIATRIC MEDICINE, KANCHI KAMAKOTI CHILD TRUST HOSPITAL.

DR.T.RAVIKUMAR, SENIOR CONSULTANT IN PEDIATRIC MEDICINE, KANCHI KAMAKOTI CHILD TRUST HOSPITAL.

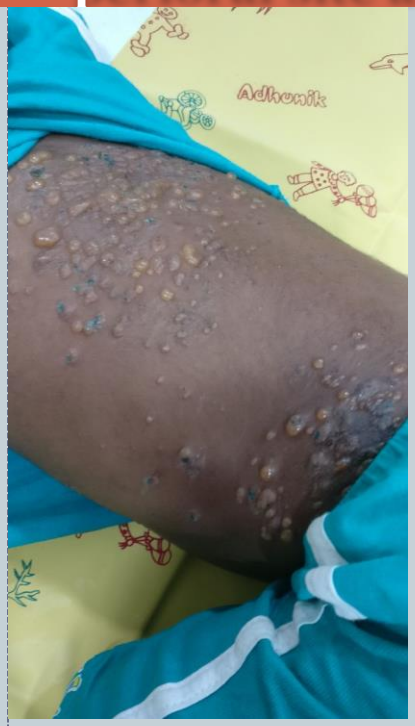
DR. RAMKUMAR, PEDIATRIC DERMATOLOGIST, KANCHI KAMAKOTI CHILD TRUST HOSPITAL.

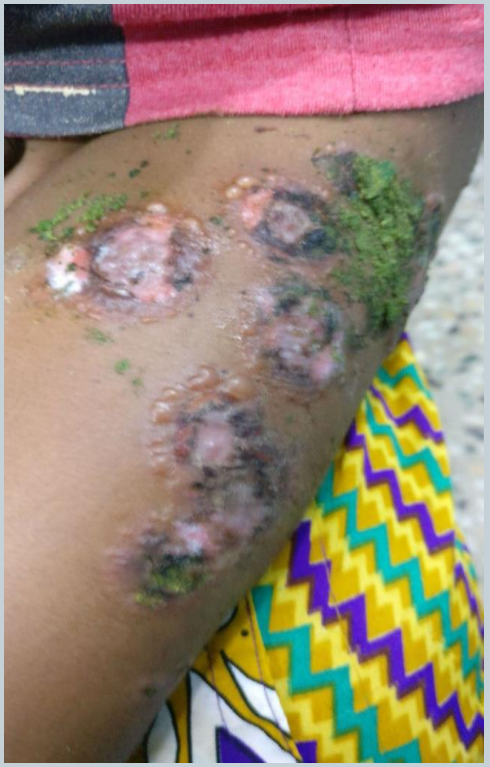
Our two cases

Hemachandar,a 2year old male child, from nellore. Rash started in lower limbs



Pranav , 2yr 8m old male child, from Chennai. Rash started in perioral site and trunk.





Background history

- First born out of non consanguineous marriage.
- With no significant past medical illness.
- No family history suggestive of genetic disorders.
- No history of preceding drug intake or no symptoms and signs suggestive viral illness.

How did lesions start

- Started initially as maculopapular rash
- Progressed to vesicles and tense bullous lesions which spread over to rest of body.
- Sparing of flexures and mucosal surfaces.
- Associated with itching from the very beginning of onset.
- Serous discharge from broken and healing bullous lesions.
- Skin lesions of different age of onset.

Other clinical features

- No fever or generalized malaise.
- No other systemic symptoms suggestive of other organ system involvement.
- These children were a little irritable initially due to pruritis , which subsided with antihistaminics coverage.
- Both had good appetite and were interested in play and surroundings
- No lesions in mucosal areas
- Absent Nikolsky's sign

WHAT DOES VESICOBULLOUS LESIONS IN A CHILD MEAN?

Infective causes	Congenital	Immunological
Staphylococcal scalded skin syndrome	Epidermolysis bullosa simplex	Pemphigus
Erythema multiforme	Junctional Epidermolysis bullosa	TEN / SJS
Bullous impetigo	Dystrophic epidermolysis bullosa	Dermatitis herpetiformis
		Chronic bullous disease of childhood

Clinical clues to pick the right disease

- Onset in early neonatal period in form of blistering at acral prominences.

→ "Epidermolysis Bullosa disorders" also called as mechanobullous disorders

- Prodromal symptoms of fever, malaise, flu like respiratory illness, skin tenderness and erythema present and have more debilitating outcome.

→ TEN/SJS

SSSS

Pemphigus Vulgaris

Clinical clues to pick the right disease

• Minimal or no prodromal symptoms	Dermatitis herpetiformis Chronic bullous disease of childhood
• Positive Nikolsky's sign	TEN Pemphigus SSSS
• Extensive mucosal involvement with severe sequelae	TEN Pemphigus

Most probable clinical diagnosis of these kids

"Immunologically mediated vesicobullous disease".... Most probably "Chronic bullous disease of childhood"..... But why??

- Onset of disease in early childhood
- Distribution of lesions in perioral areas, hands and feet, lower abdomen and perineum.
- Lack of severe prodromal symptoms.
- Playful child inspite of dangerous looking skin lesions.
- Tense bullous lesions.

How did we prove it with minimal investigations??

Histopathological examination of full thickness skin biopsy specimen:

- Focal basal vacuolation and sub epidermal bullae.
- Neutrophilic micro abscess in dermal papillae.

Direct immunofluorescence of skin biopsy specimen:

- Linear deposits of IgA and mild deposits of IgG and C3 at dermoepidermal junction

TREATMENT OF CHOICE

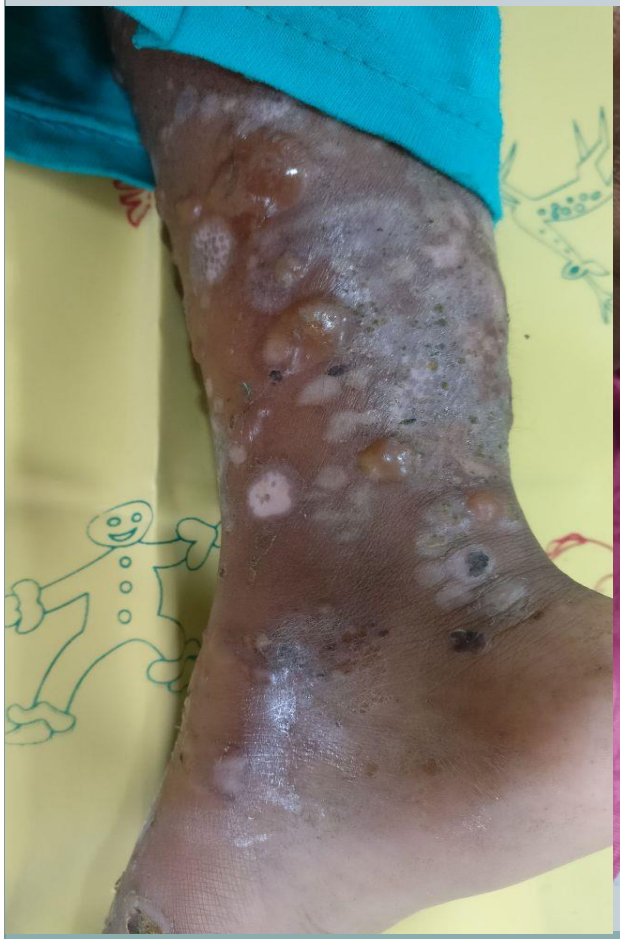
DAPSONE- FIRST DRUG OF CHOICE

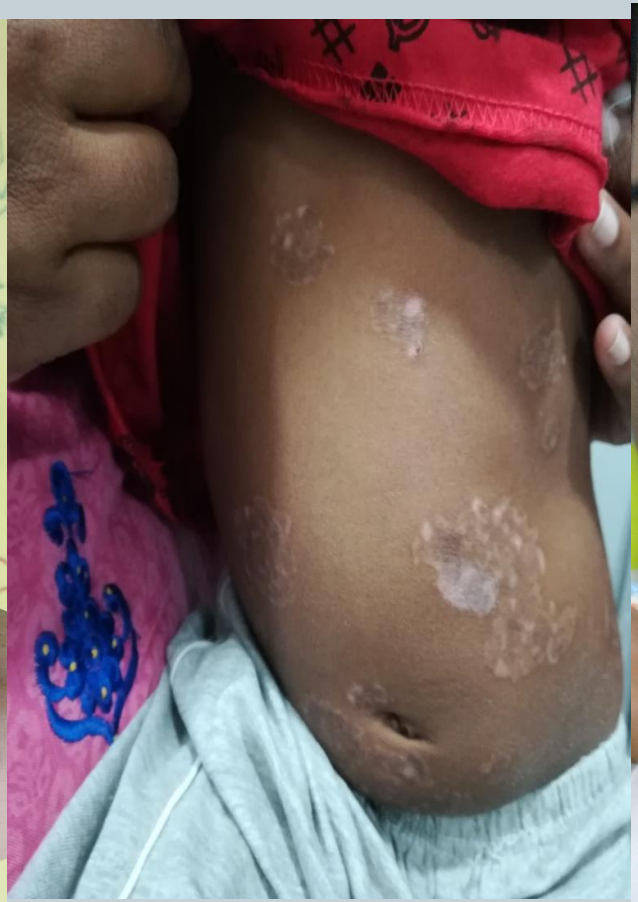
- Dose -2mg/kg/day daily till resolution of lesions.
- Must rule out severe anemia and liver dysfunction.
- Must rule out G6PD and pyruvate kinase deficiency.

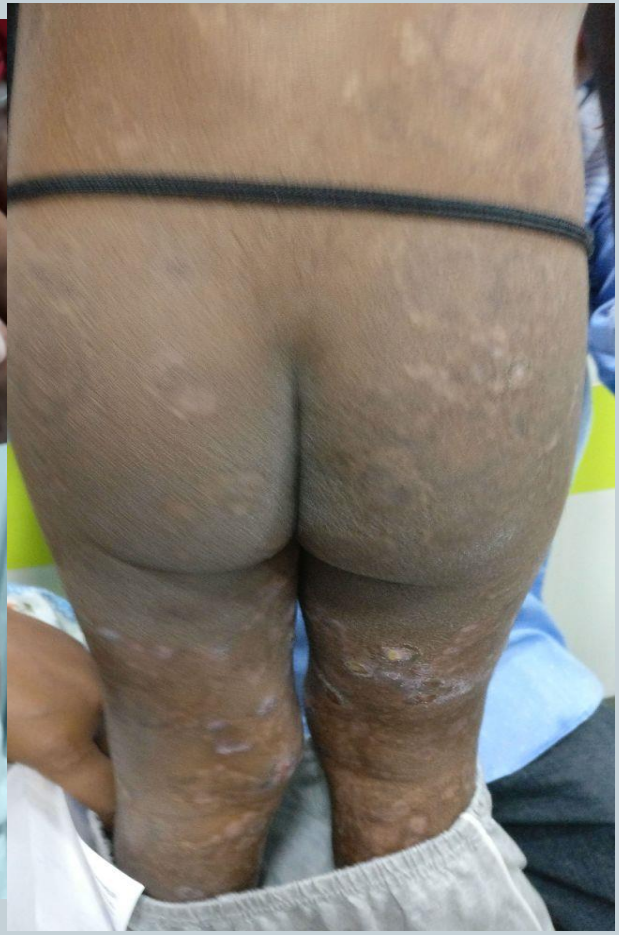
"One of our two cases proven to be G6PD deficient….
Hence this child was started on **cyclosporine**."

Steroids- are only used as add on therapy as relapse rates are very high.

Outcome of child on Dapsone therapy







To summarise….

- Chronic bullous disease of childhood, also known as linear IgA dermatosis, is an autoimmune sub epidermal vesiculobullous disorder.
- This may be idiopathic or drug induced. Vancomycin – most commonly implicated dug.
- Bimodal age distribution. In children- 6months to 10years.
- Better remission rates in children; mostly remit in 2 years.

To summarise…

- Cutaneous lesions distribution sites- lower abdomen, anogenital area, face mainly in perioral site, feet and hands.
- Also called cluster of jewel appearance.
- Cutaneous lesions heal without scarring.
- Mucosal surfaces with stratified squamous epithelium mostly affected- Oral and ocular sites.
- Minimal prodromal symptoms such as transient pruritis and no systemic toxic features.

To summarise

- Association with autoimmune lymphoproliferative disease, ulcerative colitis and SLE have been reported in adults.
- Direct immunofluorescence is investigation of choice.
- Dapsone is first drug of choice.
- Steroids, sulphapyrdine, colchicine and tetracyclines also used for treatment

THANK YOU

References…

- Jayakar Thomas. Pediatric dermatology wards. Jaypee publication. First edition. 2007.
- Arun C Inamdar. Textbook of pediatric dermatology. Jaypee publication. 2nd edition.
- Seanchen; Peter Mattei. Linear IgA bullous dermatosis. Journal: eplasty. Pubmed.
- PastuszczakM; LipkogodlewskaS. Drug induced linear IgA bullous dermatosis after discontinuation of cefuroxime axetil treatment. Journal dermatology case report. Dec2012.