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APLASIA CUTIS CONGENITA IN A NEWBORN ASSOCIATED TO VARICELLA VIRUS INFECTION DURING PREGNANCY

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Aplasia cutis congenita (ACC) is a rare skin condition¹⁻² characterized by complete or partial absence of the layers of the skin that usually presents as a unique skin lesion frequently localized in the scalp with only 20%¹ of cases presenting in the extremities. We present the case of a newborn affected by ACC localized in the left upper extremity associated to intrauterine varicella infection.

We present the case of a male newborn, son to a 19 year-old mother without significant medical history, G1, reporting 7 prenatal care appointments with folic acid intake. The mother reported diagnosis of varicella virus infection during the 2nd trimester of pregnancy (16.4 weeks of gestation) by clinical evidence of varicella scars at time of pregnancy culmination. The mother denied receiving treatment. No other prenatal conditions were declared. The patient was born at term (37.4 weeks) through vaginal delivery with use of forceps, APGAR 8/9, weighing 3.220 kg (50-90th%), 51 cm long (90th%). During the physical examination 3 skin lesions were identified, the first localized in the medial region of the left forearm extending in a lineal direction from the cubital fossa to the wrist consisting on an erythematous plaque (7 × 2 cm) with confluent pink papules, with areas of ulceration and necrosis covered by a thin membrane. The second lesion of the same characteristics was localized in the left axillary fossa (1 × 0.5 cm) and a third ulcerative lesion in the left interdigital fossa between the 4th and 5th finger with associated hypoplasia and anonychia of the latter.

Conclusion: Type VIII ACC was diagnosed according to the Frieden Classification³⁻⁴ which is attributed to teratogens and intrauterine infections including varicella virus. Limb anomalies raised the index of suspicion for Fetal Varicella Syndrome (FVS) an infrequent complication occurring only in 2% of newborns with intrauterine infections with Varicella during weeks 7-28 of pregnancy^{4, 5} but no other anomalies were found in our patient associated to FVS, the patient will be followed up for neurological sequelae. This case is a rare presentation of ACC due to the association to intrauterine varicella infection, exposure to teratogens during pregnancy should be actively investigated in ACC.

¹Martinez-Guisasola, J, Guerrero Ibanez, M, et al. Aplasia cutis congenita. Prenatal diagnosis. Elsevier. *Prog Obstet Ginecol* 2010; 53:485-9 - <https://doi.org/10.1016/j.pog.2010.05.004>.

²Knopfel, N. (2014). Aplasia cutis congenita: Etiopathogenesis, diagnostic approach and therapeutic management. Elsevier. *Piel*. 2015; 30 (2):102– 112.

³Mesrati, H. Amouri, M. (2015). Aplasia cutis congenita: report of 22 cases. *International Journal of Dermatology* 2015, 54, 1370–1375.

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⁵Sharma, S. Sharma, V. et al. Varicella Fetal Syndrome – A case report. *Curr Pediatr Res* 2012; 16 (1): 5-8.

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CONGENITAL EROSIIVE AND VESICULAR DERMATOSIS HEALING WITH RETICULATED SUPPLE SCARRING: REPORT OF FOUR CASES

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Congenital erosive vesicular dermatosis (CEVD) is a rare disorder that presents with erosions at birth and heals with reticulated supple scarring. Most of the neonates born are premature with some patients showing neurological involvement, scarring alopecias, hypohidrosis and hypoplastic nails. Although the etiopathogenesis of this entity remains unknown multiple hypothesis have been proposed. Although clinically distinct it does pose a diagnostic dilemma to the treating physician owing to confusion with other differential diagnosis like herpetic infections, epidermolysis bullosa, congenital ichthyosis in the neonates and other conditions like Kindler syndrome and porphyrias in the school going age group where it may present only with scarring. The current case presents 2 siblings born of third degree consanguineous marriage who presented with only scarring to the dermatology outpatient department. A thorough evaluation with respect to clinical history, examination and investigations- histopathology, urine porphyrin levels and antigen mapping was done to arrive at the diagnosis of CEVD.

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CASE OF PEMPHIGOID GESTATIONIS IN A MOTHER AND NEWBORN

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The 6-days-old girl was consulted at the emergency department with skin lesions on the face, trunk and extremities, which had appeared on the third day of life. The girl was born by Cesarean section at the age of 35-36 weeks because of mother's skin disease. During the postnatal period no fever occurred. Physical examination revealed multiple erythematous plaques scattered all over the body, onto her face we observed several blisters containing serous fluid, some of them were dried up and crusts were formed. Provisional diagnosis Pemphigoid gestationis was established. Lab investigation detected BP180 autoantibodies, confirming the provisional diagnosis. The mother suffered from pruritic skin lesions with redness on the abdomen, which appeared at the 30th week of her gestation, at the 34th week of gestation mother developed multiple erythematous plaques and bullae that spread out over the entire surface of her trunk and extremities. The skin lesions found in the baby healed spontaneously during 3 weeks

without further treatment. The mother was treated by oral prednisone 60 mg with slow tapering of dosage, in a week the blisters and the erythema disappeared and no new blisters were detected.

Discussion: The pemphigoid gestationis is a rare bullous autoimmune disease occurring in approximately 1 of 50 000 pregnancies and which in 5-10% of the cases also manifests in the newborn baby¹. It characteristically appears during the second or third trimester of pregnancy². Most patients develop antibodies against hemidesmosomal proteins BP180 and less frequently BP280³. Pemphigoid gestationis typically manifests with an abrupt onset of extremely pruritic urticarial papules and blisters on the abdomen and trunk. The diagnosis is clinical but may need confirmation with skin biopsy to differentiate it from other dermatological conditions. Approximately 10% of all neonates of mothers with pemphigoid gestationis have cutaneous lesions similar to pemphigoid gestationis⁴. The disease in children seems to originate from passive transfer of the pemphigoid antibodies, but could also be caused by resting maternal hormones⁵. The skin lesions resolve spontaneously in the first months and in most of the cases treatment of the child is not necessary.

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GENERALIZED CUTIS MARMORATA TELANGIECTATICA CONGENITA: A CASE REPORT

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Cutis Marmorata Telangiectatica Congenita (CMTC) is a rare cutaneous vascular malformation compromising venous and capillary vessels, clinically characterized by the presence of erythematopurpural maculae forming a reticular pattern which usually is localized in the lower limbs, but in rare cases can be generalized. The etiology of CMTC is unknown and it has an incidence of 1/3,000 births without sex predominance. The diagnosis is made base on clinical basis, since the histopathological features are non specific. CMTC has a good prognosis but it depends on the associated malformations, which are present in 20-80% of cases. We present the case of a 45 day old male with generalized lesions compatible with CMTC that were present since birth. The patient had left pielectical system dilation and right hydrocele as associated malformations.

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INDUCTION OF EPIDERMAL DAMAGE BY TAPE STRIPPING TO EVALUATE NEW CLEANSING REGIMENS FOR THE PREMATURE EPIDERMAL BARRIER

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Previous work and publications indicate a compromised skin barrier model may be a possible surrogate for premature, undeveloped skin. This study was done to confirm the consistency of the model as a surrogate test design and to test baby wipes for mildness and gentleness.

This study involved serial tape stripping the volar forearms of adult female volunteers (n=34) to "thin" the stratum corneum. The forearm sites received intensive applications of different wiping options (two wipes, washcloth & water, and no wipe control) that exaggerated the cleansing exposures beyond what a child in the NICU might experience. The recovery of skin barrier function during the wiping regimen, measured by trans-epidermal water loss (TEWL), was used as the primary indicator of mildness of the cleansing options. TEWL measurements were made prior to the first wash on days 1-4, on day 5 (primary endpoint), and at the final visit on day 8. Erythema grading was done as a secondary endpoint.

Epidermal barrier function was assessed using trans-epidermal water loss (TEWL) as the primary endpoint with skin redness (erythema) as a secondary measure. The Pampers wipes were not statistically significantly different ($p = 0.4853$) from each other for TEWL on Day 5. The new Pampers wipes were statistically superior ($p=0.0096$) to wash cloth and water (-6.76 g/m²/hr). The new Pampers wipes had a statistically significantly higher ($p < 0.0001$) Day 5 TEWL mean (26.36 g/m²/hr) than the tape stripped control site (15.98 g/m²/hr) but this was consistent with the currently marketed wipe (25.07 g/m²/hr). Days 1-4 were not compared. The Day 8 measurements show that after cessation of the wiping, the washcloth and water treatment recovered to a similar level as the other treatments, confirming that the wiping was inhibiting skin barrier recovery. Erythema (redness) grading shows similar trends as the TEWL results.

The study confirms that this model shows promise as a possible surrogate model for assessing skin mildness of cleansing products that may be used in the care of premature infants, and that current baby wipes products are consistently gentler to the skin than cotton washcloth and water.

- O'Connor, Robert J., et al. "Induction of epidermal damage by tape stripping to evaluate skin mildness of cleansing regimens for the premature epidermal barrier." *International Journal of Dermatology* 2016, 55 (Suppl. 1): 21–27.