

# IADV L

**SIG Pediatric Dermatology (IADV L Academy) Newsletter**

**Issue 4 , December 2017**



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# Pearls in Pediatric Dermatology

Dr. V. Anandan

1. Any maculopapular rash- don't forget to look at palms, soles and mucous membranes as it denotes more serious rash and also possibly drug reaction.
2. Any midline lesion – rule out CNS involvement as it may suggest embryonic malformation.
3. Itch is an essential symptom for Atopic dermatitis.
4. Progeric changes of hair and skin in children is a marker for ataxia telengectasia and usually has ocular involvement in the form of abnormal eye movements.
5. In case of unilateral port wine stain always rule out Sturge Weber syndrome and in case of midline port wine stain look out for neural tube defects.
6. Segmental or plaque facial hemangioma requires evaluation to rule out PHACES syndrome (Posterior fossa malformation, facial Hemangioma, Arterial anomalies, Cardiac defects and Eye and Sternal abnormalities).
7. Hyperpigmentation of knuckles, mucocutaneous membranes and unresolving skin lesions is a red flag sign for vitamin B12 deficiency.
8. Diffuse papulosquamous scaly lesions with perioral, perianal and acral involvement necessitates evaluation of zinc deficiency- Acrodermatitis Enteropathica.
9. Failure to thrive with recurrent skin infections like abscess, folliculitis and furuncles along with respiratory infection requires evaluation for Hyper IgE syndrome.
10. Eczematous rash with thrombocytopenia – consider Wiskott Aldrich syndrome; Silver hair refractory to therapy – consider Chédiak–Higashi syndrome; Both have bacterial infections common to them.
11. Giant Molluscum contagiosum with extensive involvement of face in children requires testing for HIV to rule out immunodeficiency.
12. Acanthosis nigricans in children should be followed up for possible development of hyperinsulinemia and diabetes mellitus later in the life.
13. Acute urticaria in children is mostly due to underlying viral infection and needs only supportive management.
14. Guttate psoriasis in children has a likely infective etiology most often due to streptococcus and can respond well to antibiotics.
15. Periungual desquamation in children is one of the markers of Kawasaki disease.
16. Hair Collar sign - a ring of high density, dark coarse and long hair seen around a smooth nodule, mostly over scalp is a marker for cranial dysraphism including meningocele, encephalocele and hypertrophic brain tissue.
17. Rash followed by occurrence of fever and disappearance of rash with fever is an indicator of roseola infantum or sixth day disease caused by human herpes virus 6 (HHV 6).
18. Friar Tuck sign which indicates loss of hair over the vertex and sparing of occipital region necessitates eliciting history of trichotillomania.
19. Perifollicular hemorrhage is a marker for scurvy.
20. Do not advice bathing a child if less than 2.5 kg and hemodynamically unstable.

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# Excerpts from Journals Relevant to Pediatric Dermatology Practice

By Dr. Rahul Mahajan and Dr. Shivani Moudgil

## ANTI INTERLEUKIN 31 RECEPTOR A ANTIBODY FOR ATOPIC DERMATITIS

Thomas Ruzicka, M.D., Jon M. Hanifin, M.D., Masutaka Furue, M.D., Ph.D., Grazyna Pulka, M.D., Izabela Mlynarczyk, M.D., Andreas Wollenberg, M.D., Ryszard Galus, M.D., Ph.D., Takafumi Etoh, M.D., Ryosuke Mihara, M.S., Hiroki Yoshida, M.S., Jonathan Stewart, M.B., Ch.B., and Kenji Kabashima, M.D., Ph.D., for the XCIMA Study Group\*

*N Engl J Med* 2017; 376:826-835 March 2, 2017 DOI: 10.1056/NEJMoa1606490

### Comments

This phase 2 trial exploring the efficacy of humanized monoclonal antibody against interleukin 31 receptor A – Nemolizumab (CIM331) in adult patients with moderate to severe atopic dermatitis (AD) inadequately controlled by topical glucocorticoids or topical calcineurin inhibitors, showed the efficacy of targeting interleukin 31 receptor in AD. It was a well-designed and well conducted randomized, double blind, placebo controlled 12 week trial designed and sponsored by Chugai Pharmaceuticals, and was adequately powered to assess the outcome. Two hundred and sixty four patients between the age group of 18-65 years underwent randomization and were divided into four groups in a 1:1:1:1 ratio to receive nemolizumab subcutaneously at a dose of 0.1 mg, 0.5 mg, or 2.0 mg per kilogram of body weight or placebo every 4 weeks or nemolizumab at a dose of 2.0 mg per kilogram every 8 weeks with placebo given at week 4 in an analysis that was exploratory in nature. At week 12, among the patients who received nemolizumab every 4 weeks, changes on the pruritus visual-analogue scale were significantly higher compared to the placebo group ( $P < 0.01$  for all comparisons). The adverse events that were associated with nemolizumab were similar

to those in the placebo group with the exception of exacerbations in atopic dermatitis and peripheral edema, which were more common among the patients receiving nemolizumab.

The merits of the study include that it provides evidence supporting the role of interleukin 31 in pathobiology of atopic dermatitis, the trial apart from the various outcome measures also assessed the effects of treatment on sleep and quality of life of the patients, provides an idea regarding the optimal dose of the drug that has the best benefit-risk profile i.e. 0.5 mg/kg every 4 weeks and additionally the researchers have also measured the anti nemolizumab antibodies during the trial period. The limitations include a relatively small sample size and short study duration, and the high dropout rate because of intermittent disease exacerbations. In addition, since only adult patients were included, the results cannot be extrapolated to the pediatric population in whom atopic eczema is much commoner. Finally, long term adverse events cannot be assessed because of the short duration and small patient sample, and the trial has not compared the efficacy of nemolizumab to other conventional therapies of atopic dermatitis. However, the trial still reports exciting results as IL-31 is increasingly being recognized as an important mediator not only in atopic eczema but also in other diseases like bullous pemphigoid and dermatitis herpetiformis.

## LONG TERM FOLLOW UP OF LYMPHATIC MALFORMATIONS IN CHILDREN TREATED WITH SILDENAFIL

Tu JH, Tafoya E, Jeng M, Teng JM

*Pediatr Dermatol.* 2017 Sep;34(5):559-565. doi: 10.1111/pde.13237.

### Comments

It is a retrospective study to evaluate long

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term clinical outcomes in pediatric lymphatic malformation patients treated with sildenafil (dose of 30 mg/day for children weighing <20 kg, and 60 mg/day for those weighing >20 kg) whose benefits in the condition were discovered serendipitously by Swetman et al. Of the 12 patients who fulfilled the inclusion criteria of the study, 10 completed the survey and reported positive therapeutic response with improvement in size and compressibility of the malformation. Parents of the children were contacted to provide a subjective response about the treatment efficacy and any long term effects of the therapy. Minimal side effects were reported that included mild flushing and priapism. The study also reported the clinical and radiological response, side effects of the therapy and additional interventions required after therapy.

The strong points of the study are that it is the first report evaluating the long term effects of sildenafil therapy in lymphatic malformations reporting a successful outcome in majority of the children with acceptable adverse effect profile. The study included clinical evaluation as well as patient and parent observations. The investigators have also hypothesized the possible mechanism of action of the drug in lymphatic malformation and thus the study underscores the potential of using sildenafil as an adjuvant therapy in the management of the condition. The limitations of the study were its small sample size, retrospective study design, subjective response assessment that has the potential of recall bias and lack of validated questionnaires and instruments to evaluate the effects of therapy. Concluding, considering the cosmetic deformity caused by large lymphatic malformations, and the relative sparsity of therapeutic modalities, sildenafil may be a useful option either as a monotherapy or in combination with other modalities like sclerotherapy.

### **TWO PHASE 3 TRIALS OF DUPILUMAB VERSUS PLACEBO IN ATOPIC DERMATITIS**

Eric L. Simpson, M.D., Thomas Bieber, M.D., Ph.D., Emma Guttman-Yassky, M.D., Ph.D., Lisa A. Beck, M.D., Andrew Blauvelt, M.D., Michael J. Cork, M.B., Ph.D., Jonathan I. Silverberg, M.D., Ph.D., M.P.H., Mette Deleuran, M.D., D.M.Sc., Yoko Kataoka, M.D., Jean-Philippe Lacour, M.D., Külli Kingo, M.D., Ph.D., Margitta Worm, M.D., Yves Poulin, M.D., Andreas Wollenberg, M.D., Yuhwen Soo, Ph.D., Neil M.H. Graham, M.B., B.S., M.D., M.P.H., Gianluca Pirozzi, M.D., Ph.D., Bolanle Akinlade, M.D., Heribert Staudinger, M.D., Ph.D., Vera Mastey, M.S., Laurent Eckert, Ph.D., Abhijit Gadkari, Ph.D., Neil Stahl,

Ph.D., George D. Yancopoulos, M.D., Ph.D., and Marius Ardeleanu, M.D., for the SOLO 1 and SOLO 2 Investigators\*

*N Engl J Med* 2016; 375:2335-2348; December 15, 2016; DOI: 10.1056/NEJMoa1610020.

### **Comments**

In these two independent, randomized, double blinded, placebo controlled, phase 3 trials - SOLO1 and SOLO2 the efficacy of dupilumab, a human monoclonal antibody against interleukin-4 receptor alpha was assessed in adult patients with moderate to severe atopic dermatitis (AD) inadequately controlled by topical treatment. Both the trials had identical design enrolling 671 patients in SOLO1 and 708 in SOLO2 who were randomly assigned in a 1:1:1 ratio to receive weekly subcutaneous injections of dupilumab (300 mg) or placebo or the same dose of dupilumab every other week alternating with placebo for 16 weeks. In SOLO 1, the primary outcome i.e. the IGA score of 0 or 1 (clear or almost clear) occurred in 85 patients (38%) who received dupilumab every other week and in 83 (37%) who received dupilumab weekly, as compared with 23 (10%) who received placebo ( $P<0.001$  for both comparisons with placebo). The results were almost similar in SOLO 2. Also EASI (EASI-75) at week 16 was reported in significantly more patients receiving dupilumab than among those receiving placebo ( $P<0.001$  for all comparisons). Dupilumab was also associated with significant reduction in pruritus score ( $P<0.001$ ) and symptoms of anxiety or depression with improvement in sleep and quality of life. The most common adverse events in the two trials were exacerbations of AD that were more in the placebo group, injection-site reactions and conjunctivitis being more common in dupilumab treated group and nasopharyngitis, the incidence of which was almost similar in both the groups.

The highlights of the study include the two trial concept to provide replication of results, comparison of scores like HADS, DLQI and POEM that indicate the symptom burden on the patients' quality of life, detailed evaluation of the adverse events and safety. Both the trials taken together were adequately powered to show the effectiveness of the new biologic. As with the Nemolizumab trial, the limitations were that the trials do not compare the efficacy of dupilumab against the conventional topical/systemic glucocorticoid or calcineurin inhibitors, the long term efficacy and safety is not addressed and inclusion of only adults as subjects, not children in

whom AD is more prevalent. Another aspect that needs to be looked at is the conflict of interest statement of the authors.

### REGENERATION OF ENTIRE HUMAN EPIDERMIS USING TRANSGENIC STEM CELLS

Tobias Hirsch, Tobias Rothoef, Norbert Teig, Johann W. Bauer, Graziella Pellegrini, Laura De Rosa, Davide Scaglione, Julia Reichelt, Alfred Klausegger, Daniela Kneisz, Oriana Romano, Alessia Secone Seconetti, Roberta Contin, Elena Enzo, Irena Jurman, Sonia Carulli, Frank Jacobsen, Thomas Luecke, Marcus Lehnhardt, Meike Fischer, Maximilian Kueckelhaus, Daniela Quaglino, Michele Morgante, Silvio Bicciat, Sergio Bondanza & Michele De Luca.

*Nature* 2017 Nov 16; 551(7680):327-332.

#### Comments

In this study the authors have demonstrated a life-saving procedure wherein they regenerated virtually the entire epidermis of a child suffering from a devastating form of junctional epidermolysis bullosa (JEB), by means of autologous transgenic keratinocyte cultures. It was performed on a seven year old child suffering from severe JEB carrying a homozygous acceptor splice site mutation within intron 14 of *LAMB3*, having almost complete epidermal loss of approximate 80% of the body surface area and an unfavorable short term prognosis. A 4 cm<sup>2</sup> biopsy sample, taken from the non-blistering area of the patient's left inguinal region, was used to establish primary keratinocyte cultures, which were then transduced with a retroviral vector expressing the full-length *LAMB3* cDNA. Sufficient (0.85 m<sup>2</sup>) transgenic epidermal grafts, enough to cover the patient's entire denuded body surface, were then applied sequentially on a properly prepared dermal wound bed. During the 21 month follow-up period the patient's epidermis was stable, did not blister, itch or required any medications. The regenerated epidermis adhered firmly to the underlying dermis which was confirmed with sequential random biopsies during follow up, and in situ hybridization using a vector-specific *t-LAMB3* probe showed that the regenerated epidermis consisted only of transgenic keratinocytes. The investigators also compared the efficacy and stability of plastic and fibrin cultured grafts that showed almost equal results, confirmed the stability of the grafts histopathologically and also showed there were no autoantibodies directed against the basement membrane zone in the patient's serum during 21 months of follow up. The study thus demonstrates

that stem cell mediated gene therapy represents a promising potential cure for this fatal inherited disorder.

By Dr. Ram Gulati

### DIAGNOSTIC ACCURACY OF PEDIATRIC TELEDERMATOLOGY USING PARENT-SUBMITTED PHOTOGRAPHS: A RANDOMIZED CLINICAL TRIAL

O'Connor DM, Jew OS, Perman MJ, Castelo-Soccio LA, Winston FK, McMahon PJ.

*JAMA Dermatol* 2017; Nov 15, doi: 10.1001/jamadermatol.2017.4280. [Epub ahead of print]

Advances in smartphone telephotography have the potential to improve access to care via direct parent-clinician interaction. However, literature is unavailable regarding the accuracy of diagnoses that apart from other factors, is quite reliant on the quality of photographs sent by the parent. Additionally, the accuracy of the diagnoses has not been formally compared with the diagnoses made in person.

The authors conducted this small study to assess whether smartphone photographs of pediatric skin conditions taken by parents were of appropriate quality to allow accurate diagnosis, and also if the diagnoses corroborated well with those made in person.

A prospective study of 40 patient-parent pairs was conducted at a pediatric dermatology clinic at Children's Hospital of Philadelphia from March 1 to September 30, 2016, to assess agreement between diagnoses made by an independent pediatric dermatologist based on in-person examination and those based on parental photographs. Half of the pairs (intervention group) were randomized for a secondary analysis to receive instructions on techniques to take good quality photographs with smartphones, while the other half formed the control group. Clinicians were blinded to whether parents had received photography instructions.

Concordance between photograph-based as compared to in-person diagnosis in the intervention versus control groups were quantified using Cohen  $\kappa$ , a measure of inter-rater agreement that takes into account the possibility of agreement occurring by chance.

Among the 40 patient-parent pairs, overall concordance between photograph-based versus in-person diagnosis was 83% (95% CI, 71%-



94%;  $\kappa=0.81$ ). Diagnostic concordance was 89% (95% CI, 75%-97%;  $\kappa=0.88$ ) in a subgroup of 37 participants with photographs considered of high enough quality to make a diagnosis. No statistically significant effect of photography technique instructions on concordance was detected (group that received instructions, 85%; group that did not receive instructions, 80%;  $P=.68$ ). Authors suggested that parent-operated smartphone photography can accurately be used as a method to provide pediatric dermatologic care.

### Comments

Considering increasing use of smartphones as telemedicine equipment, especially with the usefulness of applications like WhatsApp in transmitting good quality images almost instantaneously, the world of tele-dermatology has transformed. This study is an attempt to compare the diagnostic accuracy of in-person versus photograph based consultation. Interestingly, statistically in this small study, it is quite comparable; however, small number of patient-parent groups studied is a significant limitation. Additionally, the legal aspect of transmitting the data electronically and safety concerns of this data are important issues to be taken into account before this mode is accepted formally.

### SCREENING GUIDELINES FOR THYROID FUNCTION IN CHILDREN WITH ALOPECIA AREATA

Patel D, Li P, Bauer AJ, Castelo-Soccio L.

*JAMA Dermatol* 2017 Dec 1;153(12):1307-1310.

No consensus exists on the incidence of thyroid disease in children with alopecia areata (AA) and it's not known if a true association exists. Consequently, screening practices for thyroid dysfunction vary widely. Therefore to reduce healthcare costs, eliminate unnecessary testing and to standardize clinical practice, a single site retrospective medical chart review was done in a pediatric dermatology clinic in a tertiary centre over a period of eight years. 298 patients with age range 0-21 years with alopecia areata who underwent thyroid function tests were included.

Parameters recorded included age at diagnosis of alopecia areata, duration of disease, severity, location, and type and past medical history and family medical history of patients were also recorded. Laboratory tests recorded included TSH, free T4, free T3, thyroid peroxidase antibodies (TPO-Abs), and thyroglobulin antibodies (Tg-Abs).

Of those with thyroid screening, patterns of AA included patchy (68%), ophiasis (13%), totalis (9%), and universalis (10%). A total of 59 (20%) patients had abnormalities on thyroid testing results. In this group of patients, hypothyroidism was the most frequent finding 29 (49%), with Hashimoto thyroiditis being the most common cause (24 [41%]). Other abnormalities included hyperthyroidism secondary to Grave disease (12 [20%]) and subclinical thyroid dysfunction (7 [12%]). Age, duration of disease, pattern of alopecia, and diagnosis of autoimmune diseases had no significant association with abnormal thyroid function tests, however, a personal history of Down syndrome ( $P=.004$ ), atopy ( $P=.009$ ), and family history of thyroid disease ( $P=.001$ ) did.

Recommendation is to restrict thyroid function tests to alopecia areata patients with a history of Down syndrome, personal history of atopy, a family history of thyroid disease, or clinical findings suggestive of potential thyroid dysfunction in a patient.

### Comments

There seems to be some literature available recommending thyroid function tests in either all or some patients of alopecia areata. In this study, 20% or one-fifth participants with alopecia areata had thyroid function abnormalities. Previous studies including Kasumagic-Halilovic E 2008 and Bakry OA et al 2014 showed 16% cases with subclinical hypothyroidism and 11.4% cases with abnormal thyroid function tests respectively. Considering the above, low threshold should be kept to test thyroid functions in patients with alopecia areata.

### ASSESSMENT OF THE EFFECTIVENESS OF TOPICAL PROPRANOLOL 4% GEL FOR INFANTILE HEMANGIOMAS

Mashiah J, Kutz A, Rabia SH, Ilan EB, Goldberg I, Sprecher E, Harel A.

*Int J Dermatol* February 1, 2017; 56 (2); 148-153.

Infantile hemangiomas (IHs) are the most common vascular proliferative lesions in children. Owing of their benign nature and natural involution, the vast majority of IHs do not require any treatment. In the past few years, topical beta blockers have been reported to be an effective treatment of superficial IHs. Consequently, clinical effectiveness and safety profile of topical propranolol 4% gel for the treatment of superficial IHs was sought.

An uncontrolled retrospective study of all cases of IHs treated with topical propranolol 4% gel between 2013 and 2015 was performed and their epidemiologic, clinical, and treatment data, including effectiveness score and safety, were reviewed.

The study included 63 patients with a total of 75 IHs. Of these IHs, 43 (57.3%) showed a good response to treatment, 19 (25.3%) a partial response, and 13 (17.33%) poor or no response, thus 62 (82.6%) had good or partial response to treatment. Response to therapy could be predicted by age at treatment initiation, treatment time, thickness of the superficial component, and size of the lesions. Only two patients reported minor local side effects while no systemic adverse effects were reported. Therefore, propranolol 4% gel was found to be a safe and effective topical therapy for superficial IHs.

### Comments

While systemic propranolol has become the drug of choice for hemangiomas of significant cosmetic and/or functional concern, topical beta blockers like timolol have been tried for superficial hemangiomas. Topical therapy unlike systemic therapy circumvents the need for monitoring. Timolol maleate 0.5% gel has been used over the past few years with good results in superficial hemangiomas. Recent studies have shown insignificant systemic absorption. Propranolol 1% cream and 3% gel in the past have been shown to be effective for superficial hemangiomas. Li G 2016 and Ge J 2016 have shown good response of combined oral propranolol and topical timolol for mixed infantile hemangiomas. Propranolol 4% gel should therefore become an important part of the armamentarium both as sole treatment of superficial hemangiomas and possibly in combination with oral therapy for compound hemangiomas, though systemic absorption leading to increased side effects need to be studied beforehand.

### RELEVANCE OF CAT AND DOG SENSITIZATION BY SKIN PRICK TESTING IN CHILDHOOD ECZEMA AND ASTHMA

Hon KL, Tsang KY, Leung TF.

*Curr Pediatr Rev* 2017 Jun 14. doi: 10.2174/1573396313666170615085018. [Epub ahead of print]

Household animal dander has been implicated as aeroallergen in childhood atopic diseases. Whether families with atopic children can keep a pet has been an ever baffling question. Study investigated if skin sensitization by cat/dog dander

was associated with disease severity and quality of life in children with atopic eczema (AE).

Demographics, skin prick test (SPT) results, disease severity (Nottingham eczema severity score NESS), Children Dermatology Life Quality Index (CDLQI), blood IgE and eosinophil counts of a cohort of AE patients were reviewed.

Of the 325 AE children evaluated, personal history of asthma was lowest (20%) in the dog-dander-positive-group but highest (61%) in both-cat-and-dog-dander-positive group ( $p=0.007$ ). Unlike dust mite or food sensitization, the incidence of cat or dog sensitization was low. Binomial logistic regression ascertained that cat-dander sensitization was associated with increasing age (adjusted odds ratio [aOR], 1.056; 95% Confidence Interval [CI], 1.006 to 1.109;  $p=0.029$ ), dust-mite sensitization (aOR, 4.625; 95% CI, 1.444 to 14.815;  $p=0.010$ ), food-allergen sensitization (aOR, 2.330; 95% CI, 1.259 to 4.310;  $p=0.007$ ) and keeping-cat-ever (aOR, 7.325; 95% CI, 1.193 to 44.971;  $p=0.032$ ); whereas dog-dander sensitization was associated with dust-mite sensitization (aOR, 9.091; 95% CI, 1.148 to 71.980;  $p=0.037$ ), food-allergen sensitization (aOR, 3.568; 95% CI, 1.341 to 9.492;  $p=0.011$ ) and keeping-dog-ever (aOR, 6.809; 95% CI, 2.179 to 21.281;  $p=0.001$ ). However, neither cat nor dog sensitization were associated with asthma, allergic rhinitis, parental or sibling atopic status, disease severity or quality of life.

The study therefore does not find direct correlation between AE severity, quality of life, asthma or allergic rhinitis with skin sensitization to cats or dogs. Study suggests that sensitized patients especially those with concomitant asthma and severe symptoms may consider non-furry alternatives if they plan to have a pet. Highly sensitized individuals, especially those with asthma co-morbidity, may have to remove their pet for a trial period to determine if symptoms improve.

### Comments

The study looks at multiple associations including AE severity, quality of life, asthma and allergic rhinitis with cat and/or dog sensitization. Though study suggests that physicians advice the same to their patients, authors do suggest caution with furry animals and trial of removal of pet in highly sensitized individuals especially those with asthma. Epstein TG 2011 showed that dog ownership significantly reduced the risk for eczema at age 4 years among dog-sensitized children while cat ownership combined with cat sensitization significantly increased the risk. Lodge CJ 2012

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found no evidence that exposure to cats or dogs at birth increase the risk of allergic disease in high-risk children. Evidence from the above studies and the possibility that removal of pet may be

associated with psychological trauma for the child, removal of pets from the families of affected children should not be one of the earlier measures at controlling eczema.



# Pediatric Dermatology Quiz

Dr. Najeeba Riyaz MD, DVD, DNB, FRCP (Glasg), MRCP, SCE Derm

1. A young girl who was very fond of this vegetable was evaluated for yellowish skin discoloration. However, her LFT was normal. What is the diagnosis?



2. A young girl with a voracious appetite had these lesions. What is the diagnosis and name the clinical sign?



3. This young boy was evaluated for recurrent abdominal pain. Endoscopy showed hamartomatous polyps.

What is the pattern of inheritance of this disease?



4. This 14-year-old boy with seizure disorder was put on valproic acid. 2 weeks later, he was admitted with severe epigastric pain and this skin lesion.

What is this sign called?

What is the next blood test the clinician should order?





5. This 15-year-old boy presented with 3 years of watery diarrhea, weight loss and diffuse muscle wasting. He has multiple severely pruritic vesicular lesions on his elbows and knees; he has scratched himself to the point of bleeding on several occasions. Peripheral smear showed hypochromic microcytic anemia and Howell Jolly bodies.

What is the diagnosis?



6. A 2-year-old girl developed high grade fever and maculopapular rash. ESR was 100 and platelet count 8 lakhs. This lesion helped to diagnose this disease.

What is the lesion?

What is the diagnosis?



7. This 6-year-old boy was evaluated for easy bruisability. His hematological work up was normal. Fundus showed angioid streaks of retina.

Name this clinical sign. What is the most probable diagnosis?



8. Identify the sign shown in a patient with nephrotic syndrome.



9. Name the drug synthesized from *Streptomyces tsukubaensis*.

10. Name the enanthem of exanthem subitum.



11. Name the typical skin lesion caused by this.



12. Name the species causing this lesion.



13. What is the relevance of this in dermatology?



14. Fitzpatrick sign is seen in which disease?

15. What is the cell of origin of Toker tumor?

16. Name the clinical sign shown in this child with malnutrition.



17. What is the typical histopathological finding of this lesion?



18. This is the nail finding in a child with intractable seizures. What is it?



19. What is Hertoghe's sign? What is its synonym?

20. What is Bazin's disease?

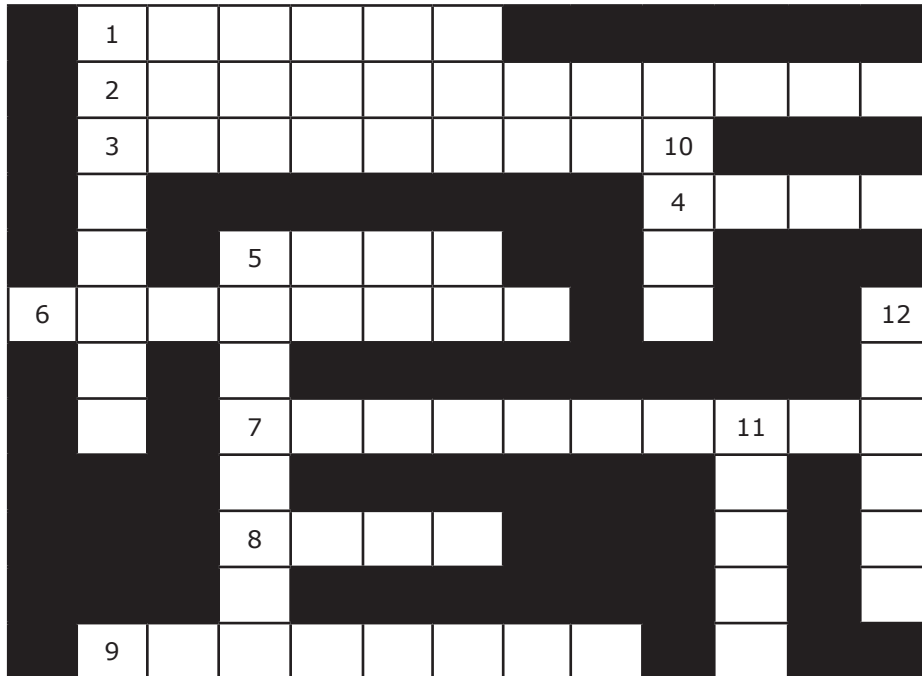


### PEDIATRIC DERMATOLOGY QUIZ- ANSWERS

1. Lycopopenia, due to consumption of large quantities of tomatoes which contain the anti-oxidant lycopene. This is similar to carotenemia. Eyes will be normal unlike jaundice.
2. Russell sign in bulimia nervosa. These children insert their fists into mouth to induce vomiting thus developing bruises in the knuckles.
3. Peutz-Jeghers syndrome- autosomal dominant.
4. This boy has developed valproic acid induced acute pancreatitis.  
Two signs have been described in hemorrhagic pancreatitis:
  - Periumbilical ecchymosis (Cullen's sign)
  - Ecchymosis of the flank (1. Grey Turner sign; 2. Amylase, lipase)
5. Dermatitis herpetiformis (Dühring's disease) in celiac disease.
6. Accentuation of BCG scar is characteristic of Kawasaki disease.
7. Gorlin sign (ability to touch nose with tip of tongue) in Ehlers Danlos syndrome.
8. Muehrcke line due to hypoalbuminemia- parallel white lines of nails. They are not grooved unlike Beau's lines.
9. Tacrolimus.
10. Nagayama spots.
11. "Breakfast, lunch and dinner rash".
12. Trichomycosis axillaris caused by *Corynebacterium*.
13. "Spaghetti and meatball" appearance in tinea versicolor.
14. Dimpling caused by squeezing the skin adjacent to a dermatofibroma is Fitzpatrick sign, also called "pinch sign" or "dimple sign".
15. Merkel cell.
16. Signe de la Bandera (flag sign) is alternate light and dark bands of hair in kwashiorkor.
17. Henderson Patterson bodies in molluscum.
18. Koenen tumor (periungual fibroma) in tuberous sclerosis.
19. Absence of lateral third of eyebrows seen in:
  - **Hypothyroidism**
  - **Hansen's disease**
  - **SLE**
  - **Atopy**
 It is also called **Queen Anne's sign**, as she had hypothyroidism.
20. Erythema induratum in TB.

# Pediatric Dermatology Crossword

Dr. Manjot



## ACROSS

- 1 - Syndrome associated with infantile hemangiomas
- 2 - A new topical antifungal agent
- 3 - mTOR receptor inhibitor
- 4 - Syndrome associated with fetal growth restrictions and gangrene of the fingers
- 5 - Common feature to both Neurofibromatosis and Tuberous Sclerosis
- 6 - One of the main constituents of stratum corneum lipids
- 7 - Hedgehog signalling inhibitor for BCC
- 8 - Roman god of all beginnings
- 9 - Lipoxygenase inhibitor used in the treatment of acne

## DOWN

- 1 - Transient Outermost layer of the epidermis
- 10 - Earliest structure of the skin to keratinize in utero
- 5 - Woolly hair with left ventricular cardiomyopathy
- 11 - Syndrome also known as Focal Dermal Hypoplasia
- 12 - Syndrome associated with Ichthyosis and Photosensitivity

## ANSWERS

### Across

1 - PHACES, 2 - Eberconazole, 3 - Rapamycin, 4 - APLA (Anti Phospholipid Antibody), 5 - CALM, 6 - Ceramide, 7 - Vismodegib, 8 - JANUS, 9 - Zileuton

### Down

1 - Periderm, 10 - Nail, 5 - Carvajal, 11 - Goltz, 12 - PIBIDS

# Upcoming Events in Pediatric Dermatology

Deepshikha Khanna

1. 43<sup>rd</sup> annual meeting of the Society for Pediatric Dermatology, Resort at Squaw Creek, Lake Tahoe, California, July 11<sup>th</sup>-14<sup>th</sup>, 2018. ([pedsderm.net](http://pedsderm.net))
2. Indian Society of Pediatric Dermatology (ISPD) Annual Conference, Goa, August 31<sup>st</sup> to September 2<sup>nd</sup>, 2018. ([peddermindia.org](http://peddermindia.org))



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