

What's new in Pediatric Dermatology

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PROGRAM

**Training Course in
Pediatric Dermatology
(SGDV/SSDV)**

Update in Pediatric Dermatology
for Dermatologist, Pediatrician and General practitioners
www.kinderhaut-zuerich.ch

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Hybrid-Event

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Management of infantile hemangiomas (IH) during the COVID pandemic

- Abrupt significant shifts in patient care (↓ ambulatory visits, ↑ telemedicine)
- Prompt initiation ↓ morbidity, ↑ long-term outcomes
- Early therapeutic intervention is critical for complicated IH to prevent medical complications or permanent disfigurement

Management of infantile hemangiomas (IH) during the COVID pandemic

- Retrospective multicenter study, n = 783, prolonged vital sign monitoring \geq 1 hour (after initial dose + dose escalation)
- Haemodynamically insignificant changes in heart rate (decrease of 8-9/min) and blood pressure (decrease of 2mm Hg)
- No symptomatic bradycardia or hypotension during monitoring
- Prolonged monitoring rarely changed management and did not predict future adverse events

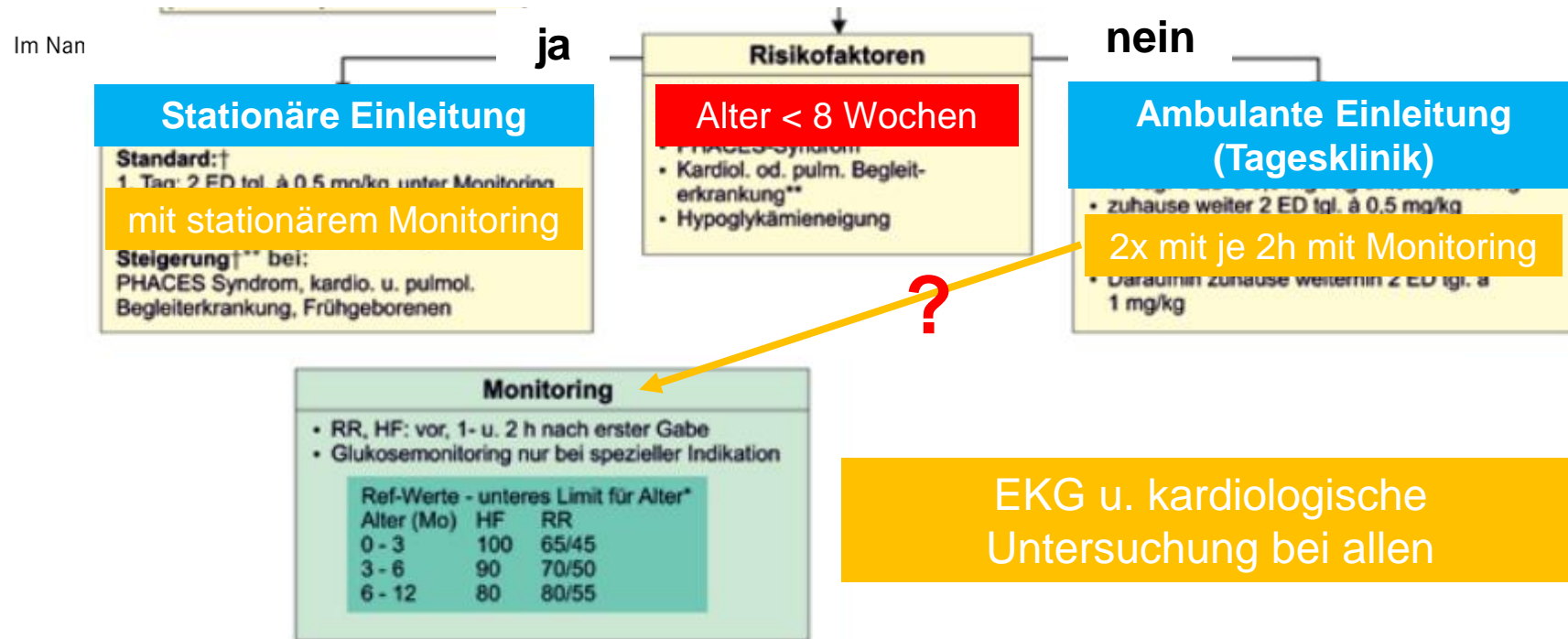
Management of infantile hemangiomas (IH) during the COVID pandemic

Consensus recommendations for management of IH by The Hemangioma Investigator Group:

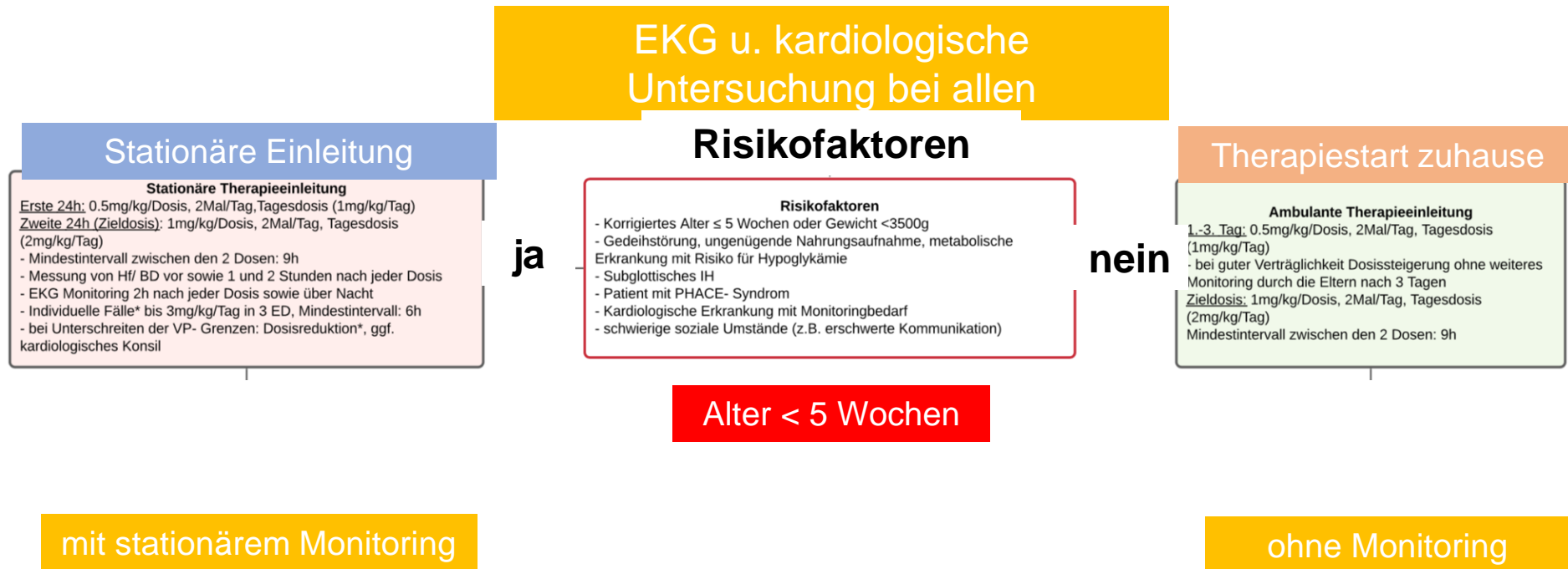
- Standard risk patients are appropriate candidates for telemedicine (oral or topical beta-blocker).
- For IH still in the rapid growth phase, particularly close follow-up is recommended.

Schweizer Richtlinien zur Propranolol-Therapie infantiler Hämangiome

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Neue Richtlinie in Bearbeitung



Kutane infantile Hämangiome (IH)



Optimal cutoff point to screen for infantile hepatic hemangioma (IHH) in patients with cutaneous infantile hemangioma (IHs)



- n = 1656 patients younger than 9 months
- Higher number of cutaneous IHs were associated with a greater risk of IHH ($p < 0.001$)
- optimal cutoff = 5 ($p < 0.001$)
- No significant association between the number of cutaneous IHs and the number of IHH ($p = 0.488$)

Screening for infantile hepatic hemangioma (IHH) in patients with cutaneous infantile hemangioma (IHs)

- Predictors of hypothyroidism in ultrasonography:
 - diffuse IHH, large (max. diameter $\geq 5.0\text{cm}$) multifocal IHH
- Recommended screening with ultrasonography in those ≥ 5 IH up to 9 months
 - exclude hypothyroidism in every IHH
- Oral propranolol treatment recommended in IHHs if
 - symptoms (hepatomegaly)
 - complications (hypothyroidism)
 - large lesion ($\geq 3.0\text{ cm}$)
 - lesion progression

Aktualisierte Liste der Geburtsgebrechen seit 1.1.2022

- Weiterentwicklung der Invalidenversicherung (WEIV) seit 1.1.2022 in Kraft
- Klare Kriterien gesetzlich festgeschrieben
- Neu IV-Arzneimittelliste (Geburtsgebrechen-Spezialitätenliste, GG-SL*)

XII. Herz, Gefäss- und Lymphsystem

311. Angeborene Hämangiome, sofern eine komplexe Therapie (mehrere Laser- oder Kryotherapien oder Operationen mit oder ohne medikamentöser Vorbehandlung) notwendig ist

How to proceed now?

- Hemangiol (on label) not on the „Spezialitätenliste“ (SL)* yet
- cost 120ml = 320 CHF (usually enough for 1 month)
- Compounded medicine (Magistralrezeptur) possible (off label)
 - Propranolol 1mg/mL in Orablend
 - (Propranolol 5mg/mL in Orablend)
- Open question: when to involve „IV“ now („complex therapy“)?



Which port-wine stain needs further investigation?

- Def. **Sturge-Weber syndrome (SWS) triad**

- port-wine stain (PWS) (Nevus flammeus)

- Ipsilateral leptomeningeal angiomas (LA)

- Oculovascular malformation (conjunctiva, episclera, choroid, retina)

- Def. **Phakomatosis pigmentovascularis (PPV) = PWS + dermal melanocytosis**

The distribution of the PWS corresponds to aberrations in embryonic vascular patterning (genetic mosaicism)

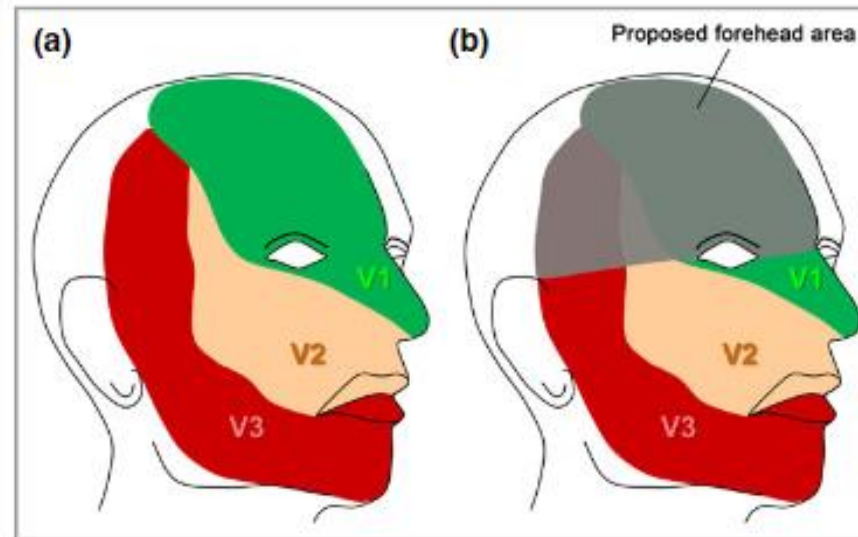


Fig 1. (a) Distribution of the three branches of the trigeminal nerve. (b) Distribution of the 'forehead', defined as any part of the forehead from the midline to an imaginary line between the outer canthus of the eye and the top of the ear including the upper eyelids. Figure adapted from *Anatomy of the Human Body*.³⁶

Forehead location and large segmental pattern of facial PWS predict risk of SWS

- More specific screening tool
- Large segmental forehead PWS* had a higher specificity and a higher positive predictive value for SWS than any forehead involvement by a PWS
- *involvement of half or more of a contiguous area of the hemiforehead (light blue) or median (purple) pattern

MRI mandatory

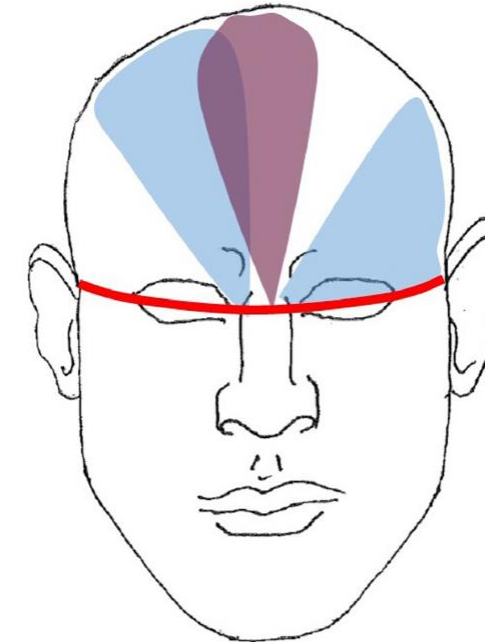


Fig 2. Forehead involvement by port-wine stains. This study defined forehead involvement as involvement by a port-wine stain of the face superior to a line connecting the outer canthus of the eye and the top of the ear, including the upper eyelid (*red line*), according to Waelchli et al.⁹ Large segmental forehead involvement was defined as involvement of half or more of a contiguous area of the hemiforehead (*light blue*) or median (*purple*) patterns. Trace or small segmental forehead involvement was defined as involvement of less than half of the hemiforehead in the absence of median forehead patterning.

Ocular manifestations of facial port-wine stain (PWS) and phakomatosis pigmentovascularis (PPV)

- Retrospective chart review of 83 patients with PWS (76 unilateral) over 10 years
- 11 with glaucoma, 4 with PWS and phakomatosis pigmentovascularis (PPV)

Ocular manifestations of facial port-wine stain (PWS) and phakomatosis pigmentovascularis (PPV)

- 18% prevalence of glaucoma in patients with PWS
- 10% will develop lat-onset glaucoma
- All patients with glaucoma presented with PWS on the eyelids

- The increase of intraocular pressure is frequently asymptomatic → irreversible damage if untreated
- Need for early and consistent monitoring of intraocular pressure

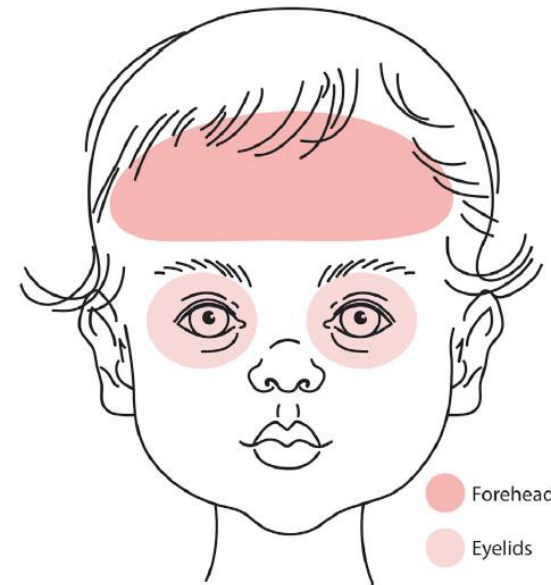


Fig 3. Facial port-wine stain. Port-wine stain involving the forehead and bilateral eyelids is associated with the highest risk of glaucoma.

No evidence of increased cancer incidence in children using topical tacrolimus for atopic dermatitis

- APPLES (A Prospective Pediatric Longitudinal Evaluation to Assess the Long-Term Safety of Tacrolimus Ointment for the Treatment of Atopic Dermatitis)
- Observational prospective cohort study, n = 7954
- Incidence over 10 years, topical tacrolimus for ≥ 6 weeks

- No lymphoma occurred



Efficacy and safety of dupilumab + top. GCS in children 6-11 years with severe atopic dermatitis

- Double-blind, 16-week, phase 3 trial
- n= 367
- 3 groups:
 - 300mg dupilumab every 4 weeks (300mg q4w)
 - weight-based every 2 weeks (< 30kg 100mg q2w, ≥ 30kg 200mg q2w)
 - placebo + medium-potency TCS

- Similar efficacy + consistent safety profile to adolescents + adults

Acne patients prescribed isotretinoin were less likely to engage in suicidal behavior under treatment

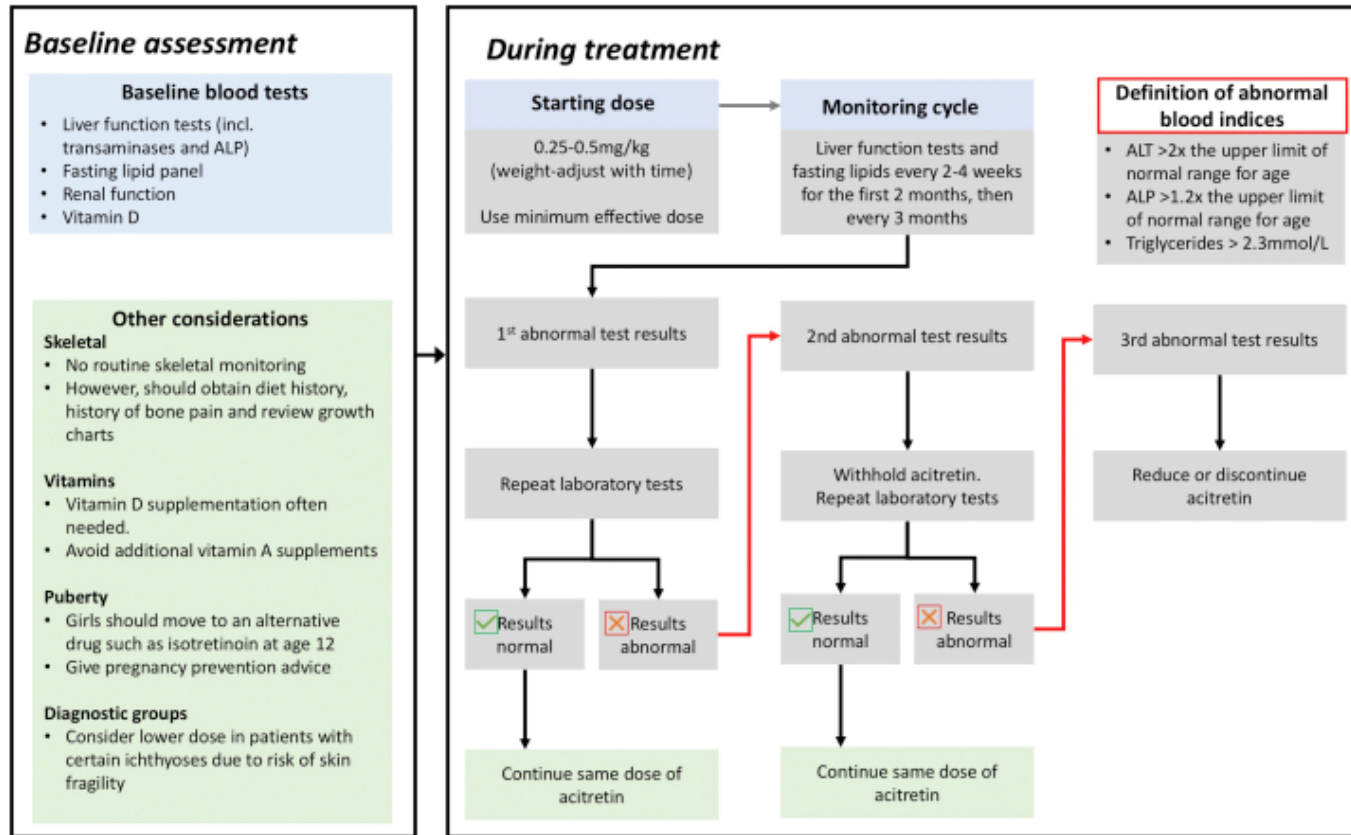
- Retrospective cohort study, n = 72'555, 12 – 35 years
- Compared to the general population, acne patients prescribed isotretinoin (or antibiotics only) were less likely to engage in suicidal behavior
- But a slight increase in suicidal behavior in isotretinoin patients 1 year after therapy

- *“We hypothesize that the increased prevalence of psychiatric diagnoses may be attributable to the impact of acne itself or increased patient contact with the health care system rather than the systemic medications used to treat acne.”*

Use of retinoids in ichthyosis and other disorders of cornification in children and adolescent

- Highly effective in reducing scaling and improving function and appearance
- Health considerations: bone, contraception
- Long-term safety data are lacking

Suggested guidelines for monitoring the use of acitretin in children

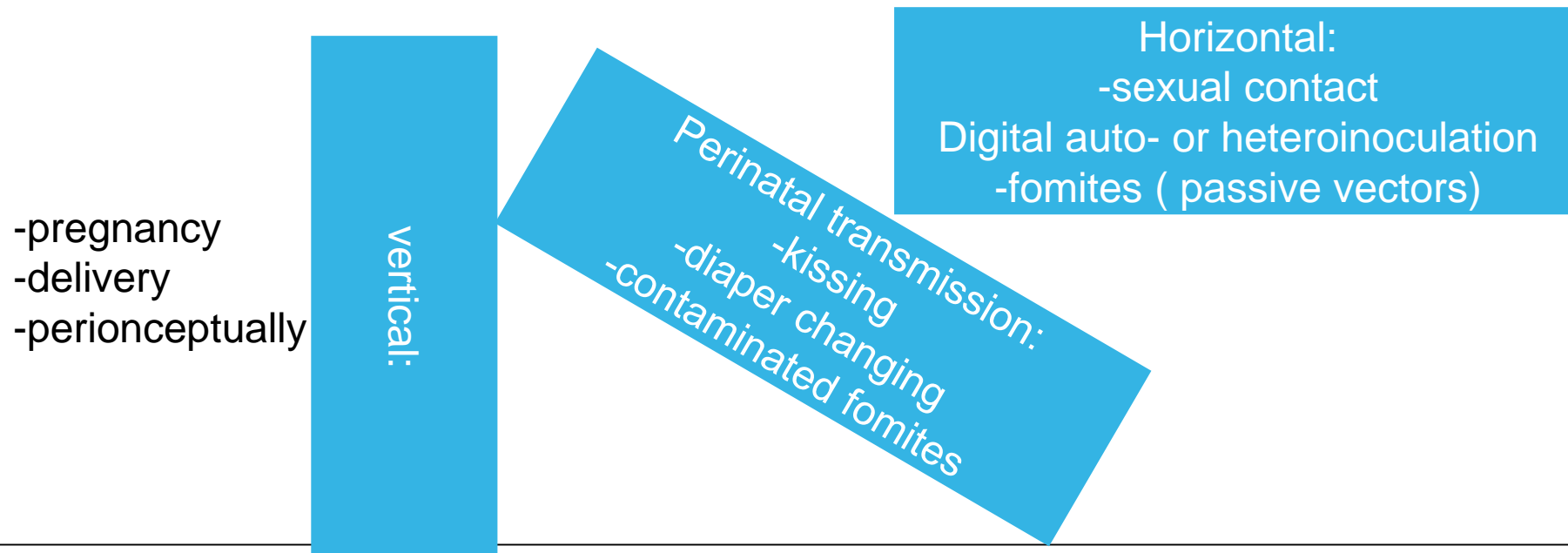


Suggested guidelines for monitoring the use of acitretin in children in association with the published guidelines in adults. *ALP*, Alkaline phosphatase; *ALT*, alanine transaminase.

Anogenital warts (AGW) and relationship to child sexual abuse?

Background:

- Human papillomavirus (HPV) is the world's most common sexually transmitted disease
- modes of transmission in children:



Anogenital warts (AGW) and relationship to child sexual abuse?

Background:

-unclear latency period of HPV (incubation period of few months to several years)

Mucosal types:

6, 11, 16, 18, 31, 33, 35,
39, 45, 52, 58, 59

Cutaneous types:

1, 4, 8, 10, 38, 41, 49

Gardasil 9 Impfung
(onkogene HPV-Typen:
16, 18, 31, 33, 45, 52, 58)

Anogenital warts (AGW) and relationship to child sexual abuse?

- Systematic review and metaanalysis, 10 studies included in the final statistical analysis (199 patients)
- Anal warts not significantly associated with sexual abuse (but genital ones)
- Overlap in HPV types found in those abused and those not abused
 - HPV typing does not confirm the mode of transmission

Take home messages

- Standard risk patients with IH are appropriate candidates for telemedicine (local and oral beta-blocker therapy)
- Investigate for IHH in 5 or more IH

- Forehead location and large segmental pattern of facial PWS predict risk of SWS
- Exclude glaucoma and monitor intraocular pressure in PWS of the forehead and bilateral eyelids

- No increased lymphoma risk under topical therapy with tacrolimus in AD
- Similar efficacy and consistent safety profile for dupilumab in children