#WCD2019

AEDV HIGHLIGHTS

24th World Congress of Dermatology (WCD)

10-15 JUNIO 2019

Milàn

Patrocina:

janssen Immunology
PHARMACEUTICAL COMPANIES OF Johnson&Johnson

Organiza:

ACADEMIA ESPAÑOLA DE DERMATOLOGÍA Y VENEREOLOGÍA
AEDV 1909

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Milán

Dermatología pediátrica

Dra. Ana Rodríguez Bandera

Patrocina: Janssen Immunology

PHARMACEUTICAL COMPANIES OF Johnson & Johnson

Organiza: AEDV Academia Española de Dermatología y Venereología 1909

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Infantile hemangiomas

Dr. Peter Hoeger

Patrocina: Janssen Immunology, Pharmaceutical Companies of Johnson & Johnson

Organiza: Academia Española de Dermatología y Venereología AEDV 1909

- Map of 4153 focal face and scalp IH
- No predilection for sites of fusion between embriologic subunits ((frontonasal, maxillary and mandibulary areas) (previous studies)
- Predilection for ocular axis, nasal tips perioral region

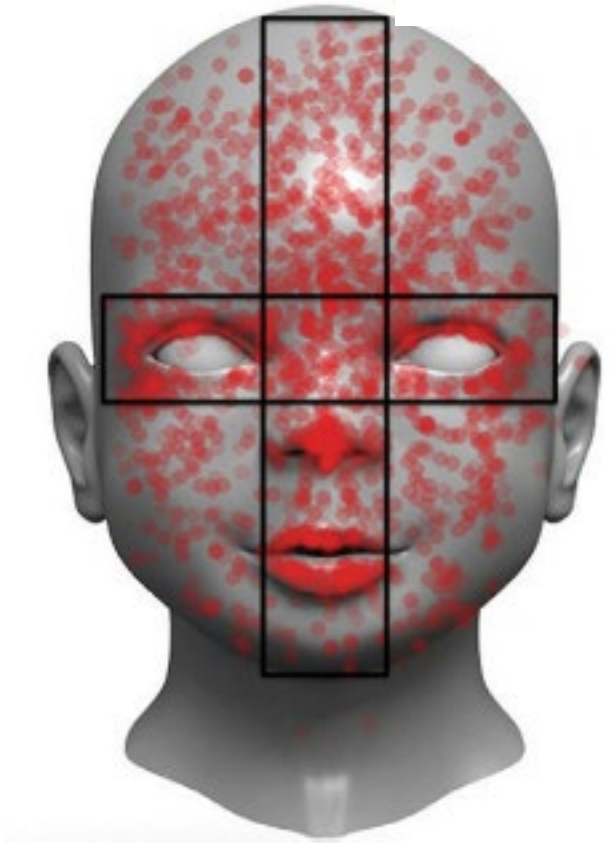

FIGURE 2 Hemangioma frequencies were greatest at the facial axis that intersected at the glabella, as shown

Haggstrom AN et al. Pediatr Dermatol. 2018

THE JOURNAL OF PEDIATRICS • www.jpeds.com

MEDICAL PROGRESS

PHACE Syndrome: Consensus-Derived Diagnosis and Care Recommendations

Maria C. Garzon, MD[1,*], Leon G. Epstein, MD[2], Geoffrey L. Heyer, MD[3,*], Peter C. Frommelt, MD[4], Darren B. Orbach, MD, PhD[5,*], Adriane L. Baylis, PhD[6,†], Francine Blei, MD[7], Patricia E. Burrows, MD[8], Sarah L. Chamlin, MD[9], Robert H. Chun, MD[10], Christopher P. Hess, MD, PhD[11,*], Shawna Joachim, BS[12,†], Katherine Johnson, DO[13], Wendy Kim, DO[14], Marilyn G. Liang, MD[15], Mohit Maheshwari, MD[8], Garrett N. McCoy, BS[12], Denise W. Metry, MD[16], Priya A. Monrad, MD[17], Elena Pope, MD[18,*], Julie Powell, MD[19], Tor A. Shwayder, MD[13], Dawn H. Siegel, MD[12,†], Megha M. Tollefson, MD[20], Sudhakar Vadivelu, DO[21], Sean M. Lew, MD[22], Ilona J. Frieden, MD[23,*], and Beth A. Drolet, MD[24,*]

Definite PHACE	
Hemangioma >5 cm in diameter of the head including scalp PLUS 1 major criteria or 2 minor criteria	Hemangioma of the neck, upper trunk or trunk and proximal upper extremity PLUS 2 major criteria

Possible PHACE		
Hemangioma > 5 cm in diameter of the head including scalp PLUS 1 minor criteria	Hemangioma of the neck, upper trunk or trunk and proximal upper extremity PLUS 1 major or 2 minor	No hemangioma PLUS 2 major criteria

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- **Objective**: Prevalence of cervical/cerebral vascular anomalies (CVA) in children with small (<5cm) vs. large (>5cm) segmental facial hemangiomas (SFH)
- **Methods**: MRI 58 patients with SFH and 50 controls
- **Results**: CVA in SFH patients: 21/58 (36%); CVA in small SFH: 6/31 (19%); CVA in large SFH (15/27; 56%); CVA in controls (5/50; (10%)
- **Conclusion**: Positive correlation between prevalence of CVA + SFH

Segmental Facial Hemangiomas
(regardless the size)
→Increased prevalence of CVA
→ MRI

Hoeger PH et al. Submitted (2019)

J Am Acad Dermatol
■ 2018

ORIGINAL ARTICLE

Late growth of infantile hemangiomas in children >3 years of age: A retrospective study

Kathleen F. O'Brien, BA, MS,[a] Sonal D. Shah, MD,[b] Elena Pope, MD, MSc,[c,d] Roderic J. Phillips, MD,[e] Francine Blei, MD, MBA,[f] Eulalia Baselga, MD,[g] Maria C. Garzon, MD,[h,i] Catherine McCuaig, MD,[j] Anita N. Haggstrom, MD,[k,l] Peter H. Hoeger, MD,[m] James R. Treat, MD,[n,o] Marissa J. Perman, MD,[n,o] Jane S. Bellet, MD,[p] Xavier Cubiró, MD,[g] Jeffrey Poole, MD,[q] and Ilona J. Frieden, MD[b]

Washington DC; San Francisco, California; Toronto, Canada; Melbourne, Australia; New York, New York; Barcelona, Spain; Montreal, Canada; Indianapolis, Indiana; Hamburg, Germany; Philadelphia, Pennsylvania; Durham, North Carolina; and New Orleans, Louisiana

Risk factors:

- Head and neck location
- Segmental morphology
- Deep dermal/subcutaneous tissue involvement

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PEDIATRICS Volume 142, number 3, September 2018

Efficacy of Propranolol Between 6 and 12 Months of Age in High-Risk Infantile Hemangioma

Eulalia Baselga, MD,[a] Bozenna Dembowska-Baginska, MD,[b] Przemysław Przewratil, MD,[c] María Antonia González-Enseñat, MD,[d] Dariusz Wyrzykowski, MD,[e] Antonio Torrelo, MD,[f] Juan-Carlos López Gutiérrez, MD,[g] Magdalena Rychłowska-Pruszyńska, MD,[h] Raúl de Lucas-Laguna, MD,[i] Altea Esteve-Martinez, MD,[j] Esther Roé, MD,[a] Mohammed Zaim, MD,[k] Yoann Menon, PhD,[k] Stéphanie Gautier, MSc,[k] Geneviève Lebbé, PhD,[l] Athmane Bouroubi, MD,[k] Alain Delarue, MD,[m] Jean-Jacques Voisard, MD[m]

- Duration of at least 6 months and up to 12 months
- **Extending treatment up to 12 months of age produced clinically meaningful increase in the success rate.**
- Treatment effect was persistent in most patients for up to 3 months without treatment
- Retreatment with propranolol was efficacious when requiered

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PEDIATRICS Volume 141, number 6, June 2018:e20173783

Safety of Oral Propranolol for Infantile Hemangioma

Catherine Droitcourt, MD,[a,b,c,d] Sandrine Kerbrat, MS,[d] Caroline Rault, PharmD,[d] Marie-Anne Botrel, DVM,[d] André Happe, PhD,[d] Ronan Garlantezec, MD, PhD,[a,e,f] Bernard Guillot, MD, PhD,[g] Jean-Marc Schleich, MD, PhD,[h] Emmanuel Oger, MD, PhD,[a,d,i] Alain Dupuy, MD, PhD[a,b,d]

- In otherwise healthy children (n=1484) increased risk of **acute bronchiolitis** (51/1484), not for bradycardia/hypotension or hypoglucemia (3/1484 each)
- In children with underlying cardiac disease (n=133), increased rate of **conduction disturbances** (11/133)
- In children with underlying respiratory disease (n=49), increased risk of **respiratory infections** (11/49)

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Hindawi
BioMed Research International
Volume 2018, Article ID 5821369, 6 pages
https://doi.org/10.1155/2018/5821369

Research Article

Effects of Propranolol on Neurodevelopmental Outcomes in Patients with Infantile Hemangioma: A Case-Control Study

Chuan Wang,[1] Qi Wang,[1] Bo Xiang,[1] Siyuan Chen,[2] Fei Xiong,[3] and Yi Ji[1]

[1]*Division of Oncology, Department of Pediatric Surgery, West China Hospital of Sichuan University, Chengdu 610041, China*
[2]*Pediatric Intensive Care Unit, Department of Critical Care Medicine, West China Hospital of Sichuan University, Chengdu 610041, China*
[3]*Department of Child Health, West China Second University Hospital, Sichuan University, Chengdu 610041, China*

Effects on neurodevelopmental outcomes in patients with IH needs further studies. Current results are conflicting.

Research letter

C. Mahon[1,2]
G. Heron[3,4]
D. Perkins[3]
A. Drage[1]
O. Wargon[3,4]

Oral propranolol for infantile haemangioma may be associated with transient gross motor delay

DOI: 10.1111/bjd.16334

Dear Editor, Oral propranolol, the first-line therapy for infantile haemangioma (IH) threatening functional impairment or formally evaluated all children with ASQ scores ≥ 2 SDs below population means.

Of 513 children treated with propranolol, 235 met inclusion criteria. Fifty-five families did not return ASQs or declined participation. In total, 162 children [110 (68%) girls; 41 (25%) < 37 weeks' gestation (range 32–37)] underwent ASQ-3 assessment and 157 ASQ:SE assessment at a mean age of 34 months (range 10–64). Premature infants aged < 2 years

Bisoprolol
Nadolol
Atenolol

- Hydrophilic
- Not crossing the BBB
- No CNS effects

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Vascular anomalies

Dra. Eulalia Baselga

Patrocina: Janssen Immunology, Pharmaceutical companies of Johnson & Johnson

Organiza: Academia Española de Dermatología y Venereología AEDV 1909

Back to overview

ISSVA classification for vascular anomalies

Type Alt ← for previous view

Simple vascular malformations I	
Capillary malformations (CM)	
Nevus simplex / salmon patch, “angel kiss”, “stork bite”	
Cutaneous and/or mucosal CM (also known as “port-wine” stain)	
Nonsyndromic CM	GNAQ
CM with CNS and/or ocular anomalies (Sturge-Weber syndrome)	GNAQ
CM with bone and/or soft tissues overgrowth	GNA11
Diffuse CM with overgrowth (DCMO)	GNA11
Reticulate CM	
CM of MIC-CAP (microcephaly-capillary malformation)	STAMBP
CM of MCAP (megalencephaly-capillary malformation-polymicrogyria)	PIK3CA
CM of CM-AVM	RASA1 / EPHB4
Cutis marmorata telangiectatica congenita (CMTC)	
Others	
Telangiectasia*	
Hereditary hemorrhagic telangiectasia (HHT) (*HHT1* ENG, *HHT2* ACVRL1, *HHT3*, *JPHT* SMAD4)	
Others	

* The CM nature of some subtypes of telangiectasia is debated.
Some telangiectasia may be reclassified in other sections in the future

Causal genes in blue

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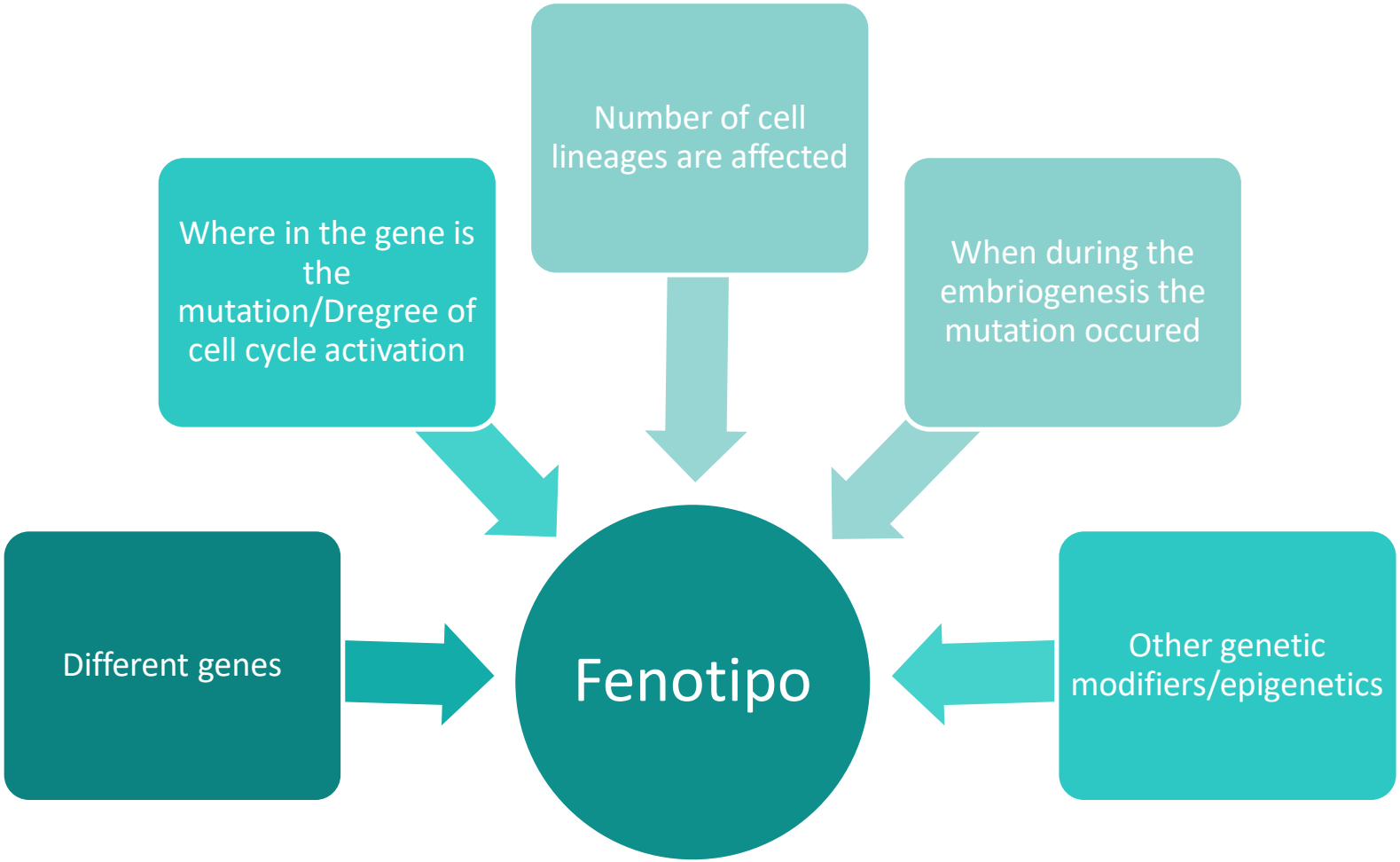
Number of cell lineages are affected
Where in the gene is the mutation/Dregree of cell cycle activation
When during the embriogenesis the mutation occured
Different genes
Fenotipo
Other genetic modifiers/epigenetics

Síndrome MC-MAV

LETTER TO THE EDITOR

Pseudocapillary malformations

RASA 1, EPHB4 mutations

Rhodoid naevus syndrome: why is this name preferable to 'capillary malformation–arteriovenous malformation'?

Retrospective analysis of 74 stains (unpublished):

- Warm, rapid capillary refill
- Sometimes minimally palpable
- Achipiellago-like borders with inlets and outlets and smaller macules "off the coast
- Not uniform in color

Dr. Baselga

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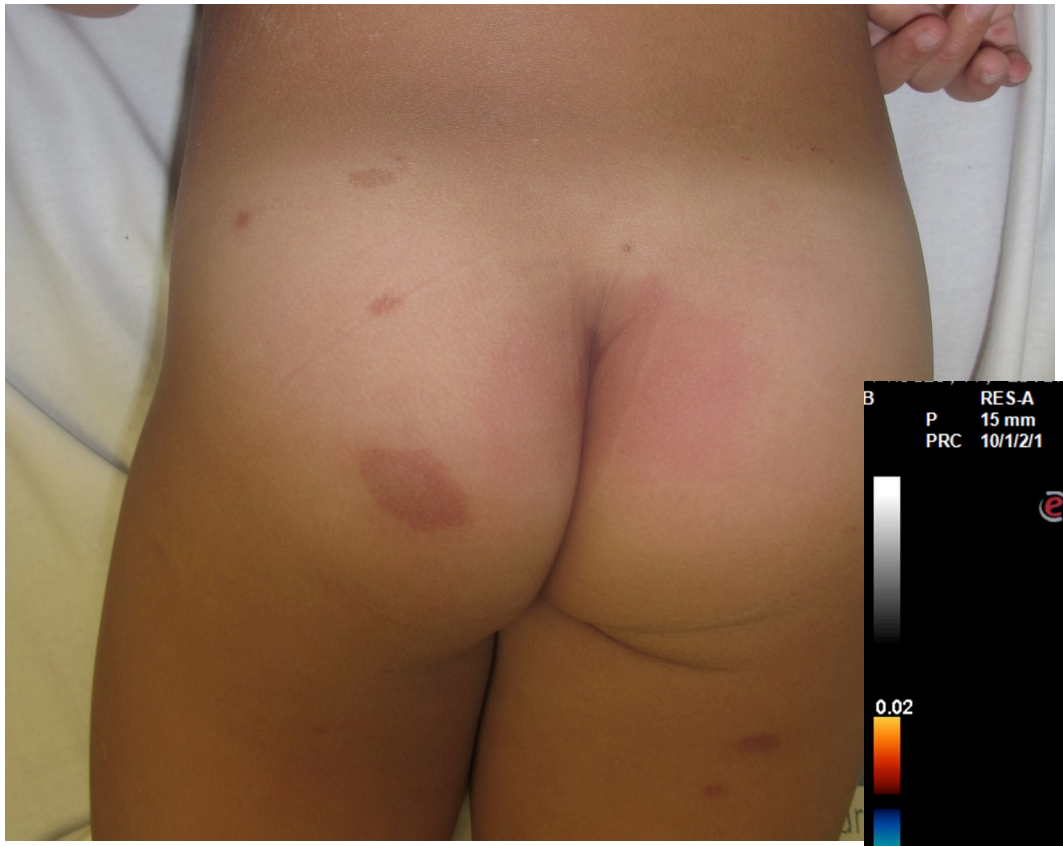

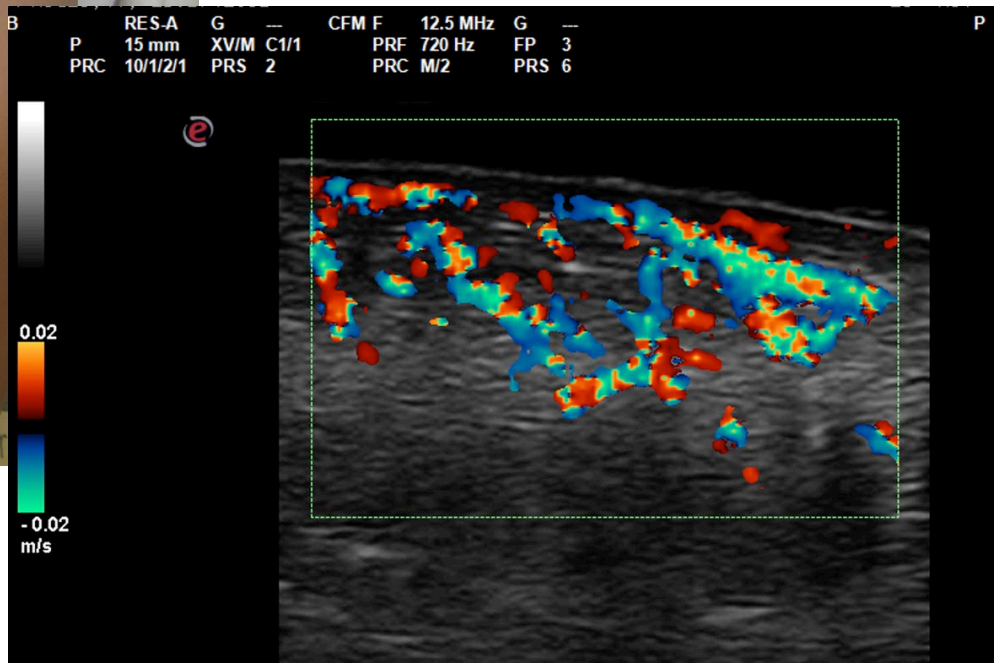

Pseudocapillary pre-AV stains

Hospital Universitario La Paz

Sirolimus (Rapamycin 0.8mg/m2/12h)

Kaposiform hemangioendothelioma
Kaposiform lymphangiomatosis
Complex lymphatic and venous malformations

Research letter

G. Zhang[1]
H. Chen[2]
Y. Gao[1]
Y. Liu[1]
J. Wang[1]
X.Y. Liu[1]

Sirolimus for treatment of Kaposiform haemangioendothelioma with Kasabach–Merritt phenomenon: a retrospective cohort study

comprehensively measured by ruler, magnetic resonance imaging or ultrasound according to the location. Treatment response was categorized into the following four groups: (i) TRR1, the tumour was reduced by ≤ 25%; (ii) TRR2, 25–50%

LYMPHATIC RESEARCH AND BIOLOGY
Volume 00, Number 00, 2019

DOI: 10.1089/lrb.2018.0068

Oral Sirolimus:
An Option in the Management of Neonates with Life-Threatening Upper Airway Lymphatic Malformations

Paloma Triana, MD,[1] Miriam Miguel, MD,[2] Mercedes Díaz, MD,[2] Marta Cabrera, MD,[3] and Juan Carlos López Gutiérrez, MD[2]

Hindawi
Case Reports in Otolaryngology
Volume 2019, Article ID 2076798, 3 pages
https://doi.org/10.1155/2019/2076798

Case Report

Sirolimus: A Successful Medical Treatment for Head and Neck Lymphatic Malformations

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