

Radiología Intervencionista en Pediatría

Nuevas estrategias

Dr. Sergio Sierre, FSIR, FCIRSE

Radiología Intervencionista

Hospital de Pediatría “Prof. J.P. Garrahan”

Buenos Aires - Argentina

1º Congreso Argentino de Medicina Interna Pediátrica

Sociedad Argentina de Pediatría

Noviembre 2016

Rx Intervencionista en Pediatría - Requisitos

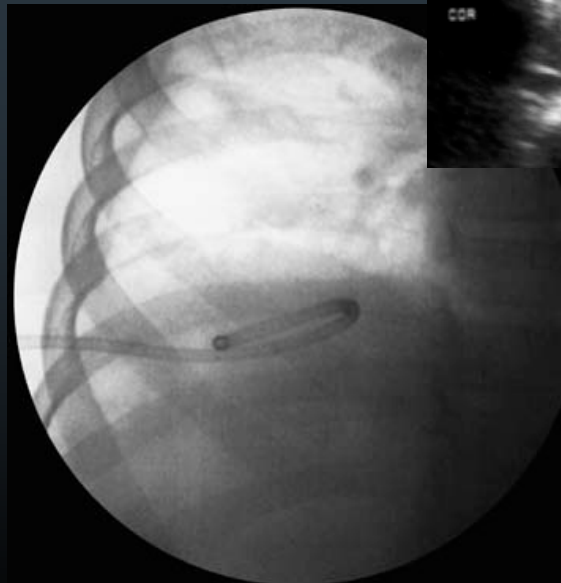
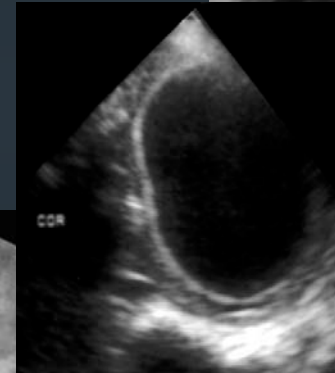
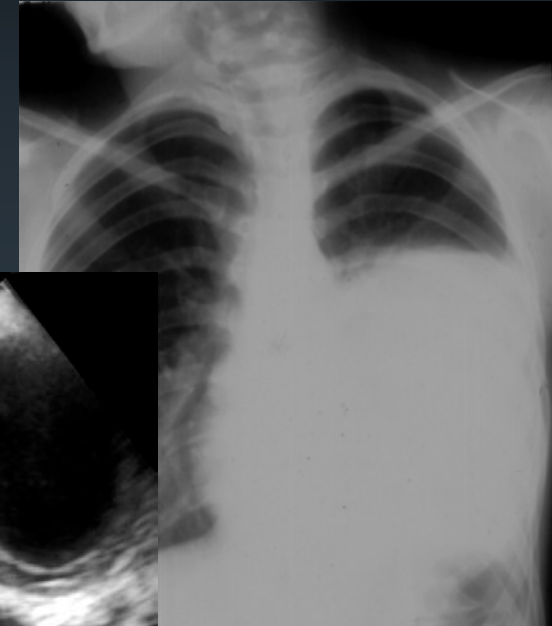
- Entrenamiento
- Conocimiento de la patología
- Adaptación de los materiales a pacientes pediátricos
- Adaptación de las técnicas
- Anestesia
- Protocolos de sedación
- Discusión, control y seguimiento multidisciplinario



Radiología Intervencionista en Pediatría

Guía por Imágenes

- Ecografía / Ecografía Doppler
- Radioscopía
- Tomografía



CLASIFICACION DE LAS ANOMALIAS VASCULARES

Mulliken and Glowacki

Mulliken JB, Glowacki J, Plast Reconstr Surg 1982 ;69:412-422

- Tumores Vasculares
- Malformaciones Vasculares

Actualización

Arch Argent Pediatr 2016;114(2):167-176 / 167

Malformaciones vasculares: actualización en diagnóstico por imágenes y tratamiento

Vascular malformations: an update on imaging and treatment

Dr. Sergio Sierra^a, Dr. Darío Teplisky^a y Dr. José Lipsich^a

Vascular Anomalies Classification and Recommendations From International Society for

TABLE 3 Classification of Vascular Tumors

Benign vascular tumors

- Infantile hemangioma/hemangioma of infancy
- Congenital hemangioma
 - Rapidly involuting CH (RICH)^a
 - Noninvoluting CH (NICH)
 - Partially involuting CH (PICH)
- Tufted angioma^{a,b}
- Spindle cell hemangioma
- Epithelioid hemangioma
- Pyogenic granuloma (or lobular capillary hemangioma)
- Others

Locally aggressive or borderline vascular tumors

- Kaposiform hemangioendothelioma^{a,b}
- Retiform hemangioendothelioma
- Papillary intralymphatic angioendothelioma, Dabska tumor
- Composite hemangioendothelioma
- Kaposi sarcoma
- Others

Malignant vascular tumors

- Angiosarcoma
- Epithelioid hemangioendothelioma
- Others

Reactive proliferative vascular lesions are listed with benign tumors.

^a Some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy.

^b Many experts believe that these are part of a spectrum rather than distinct entities.

Ahmad Alomari, MD^a, Maria C. Garzon, MD^b, Ally Mitchel, MD^c, Julie Powell, MD^d, Julie Prendiville, MD^e, Scientific Committee

ome

ations

mbined

F, AVM

M, CLM

M, CLVM

VM

AVM

ormation;

capillary-

capillary-

ic-venous

ion; LVM,

TABLE 5 Combined Vascular Malformations

Combined Vascular Malformations	
CM + VM	Capillary-venous malformation
CM + LM	Capillary-lymphatic malformation
CM + AVM	Capillary-arteriovenous malformation
LM + VM	Lymphatic-venous malformation
CM + LM + VM	Capillary-lymphatic-venous malformation
CM + LM + AVM	Capillary-lymphatic-arteriovenous malformation
CM + VM + AVM	Capillary-venous-arteriovenous malformation
CM + LM + VM + AVM	Capillary-lymphatic-venous-arteriovenous malformation

TABLE 2 2014 ISSVA Classification of Vascular Malformations

Vascular Tumors	Vascular	
	Simple	Complex
Benign	CM	See 1
Locally aggressive or borderline	LM	
	VM	
Malignant	AVM	
	Arteriovenous fistula	

TABLE 6 Vascular Malformations Associated With Other Anomalies

- Klippel-Trenaunay syndrome: CM + VM +/- LM + limb overgrowth
 - Parkes-Weber syndrome: CM + AVF + limb overgrowth
 - Servelle-Martorell syndrome: limb VM + bone undergrowth
 - Sturge-Weber syndrome: facial + leptomeningeal CM + ocular anomalies +/- bone and/or soft tissue overgrowth
 - Limb CM + congenital nonprogressive limb hypertrophy
 - Maffucci syndrome: VM +/- spindle cell hemangioma + enchondroma
 - Macrocephaly-CM (M-CM)/megalencephaly-CM-polymicrogyria (MCAP)
 - Microcephaly-CM (MICCAP)
 - CLOVES syndrome: LM + VM + CM +/- AVM + lipomatous overgrowth
 - Proteus syndrome: CM, VM and/or LM + asymmetric somatic overgrowth
 - Bannayan-Riley-Ruvalcaba syndrome: AVM + VM + macrocephaly, lipomatous overgrowth
- CLOVES, congenital, lipomatous, overgrowth, vascular

***Wassef M, et al. Pediatrics 2015; 136 (1): e203-14**

Hemangiomas

- Tumor más común en la infancia
- Superficiales, Profundos o Mixtos
- Unicos / Múltiples
- Cutáneos / Viscerales
- Aparecen en las primeras semanas de vida
- Más frecuentes en el sexo femenino (3:1)

- Localización

80 % Cabeza y Cuello

20 % Tronco y extremidades



Hemangiomas



3 meses



5 años



2 meses



9 meses

ORIGINAL ARTICLE

A Randomized, Controlled Trial of Oral Propranolol in Infantile Hemangioma

C. Léauté-Labrère, P. Hoeger, J. Mazereeuw-Hautier, L. Guibaud, E. Baselga, G. Posiunas, R.J. Phillips, H. Caceres, J.C. Lopez Gutierrez, R. Ballona, S.F. Friedlander, J. Powell, D. Perek, B. Metz, S. Barbarot, A. Maruani, Z.Z. Szalai, A. Krol, O. Boccara, R. Foelster-Holst, M.I. Febrer Bosch, J. Su, H. Buckova, A. Torrelo, F. Cambazard, B. Grzybowski, G. Wozniak, D. Wondolowski, J. Boccia, J. Bernabeu-Wi, L. Benjamin, C.R. Mehta, S. Pr

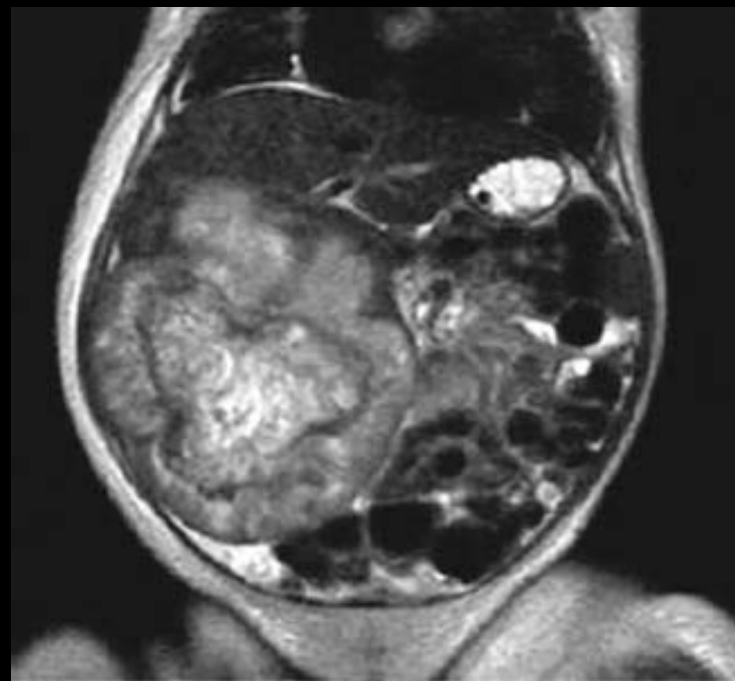
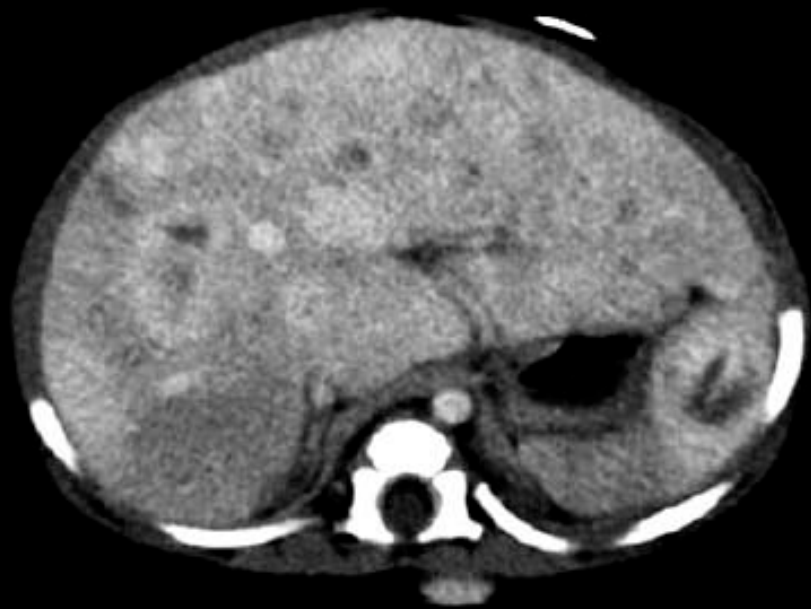
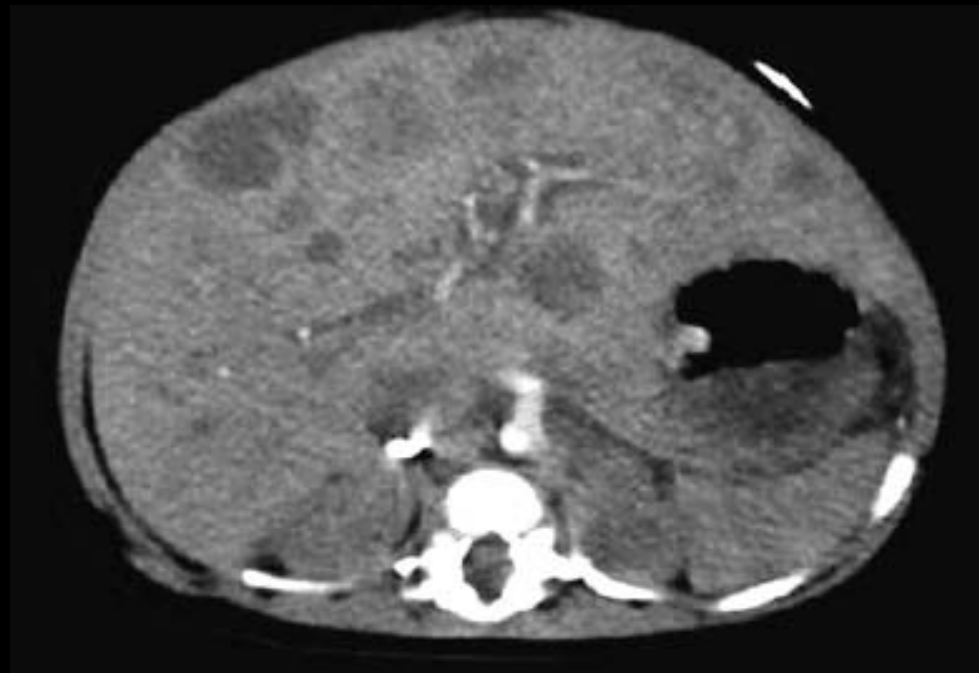
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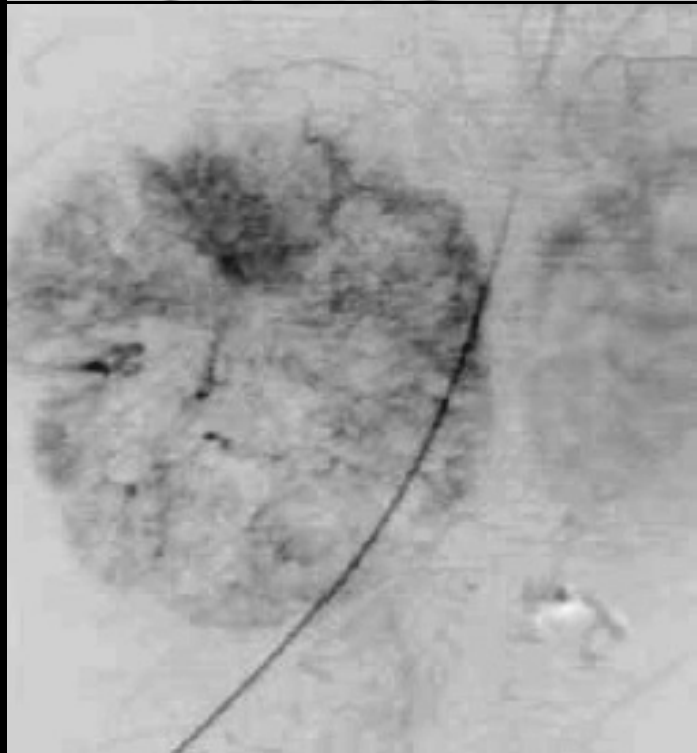
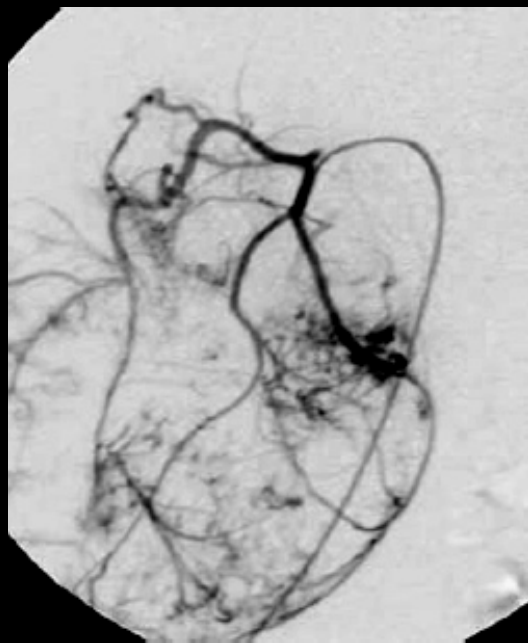
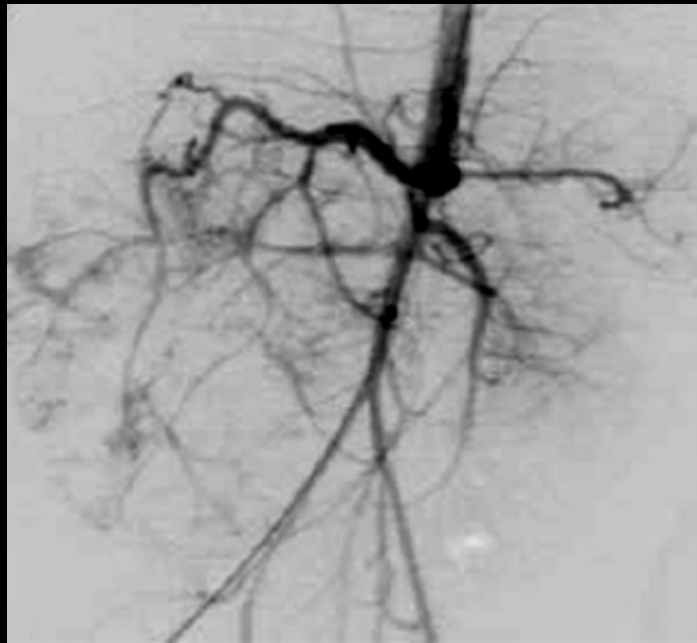
OFFICIAL JOURNAL OF THE AMERICAN ACADEMY OF PEDIATRICS

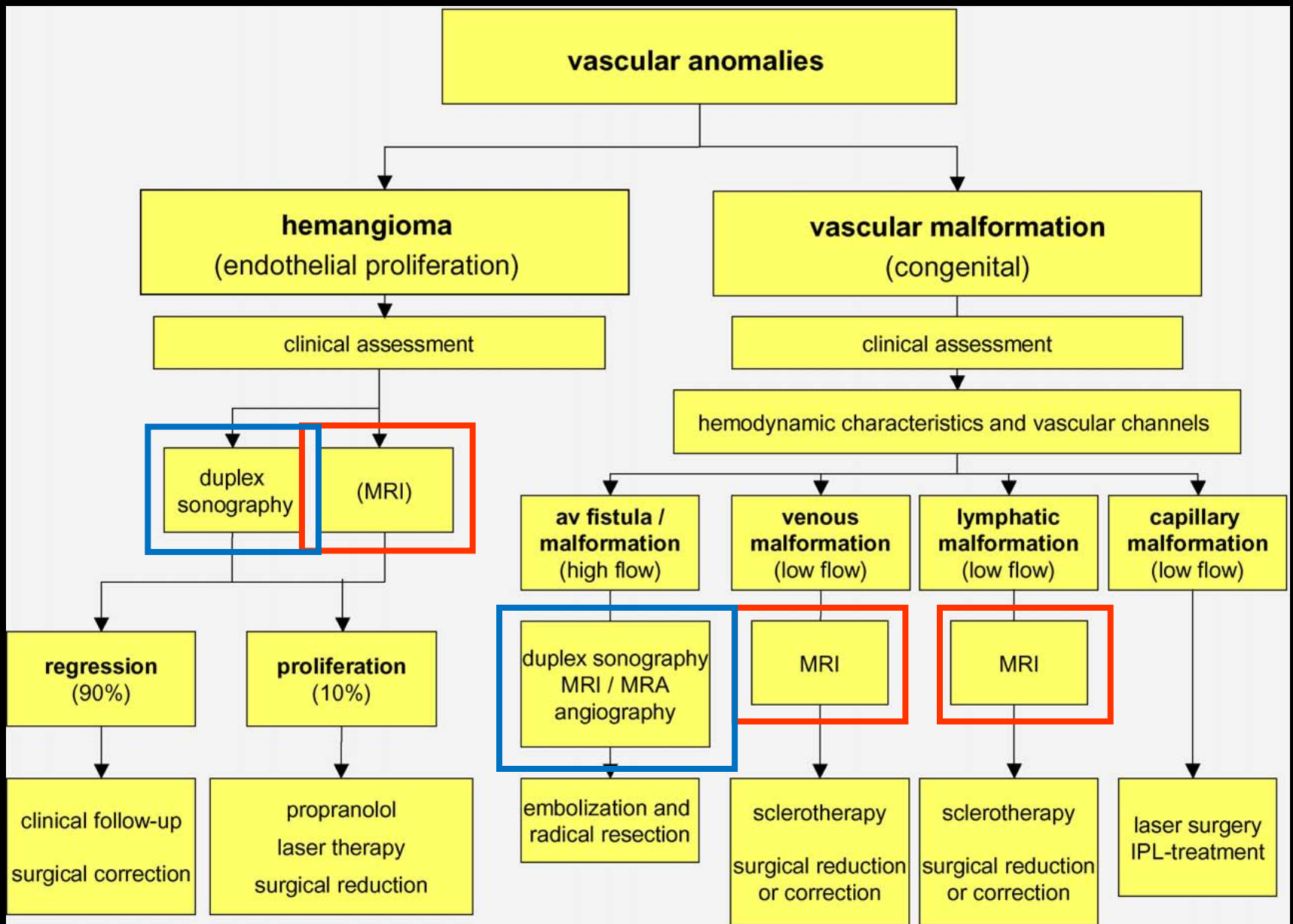
Propranolol for Severe Infantile Hemangiomas: Follow-Up Report

Véronique Sans, Eric Dumas de la Roque, Jérôme Berge, Nicolas Grenier, Franck Boralevi, Juliette Mazereeuw-Hautier, Dan Lipsker, Elisabeth Dupuis, Khaled Ezzedine, Pierre Vergnes, Alain Taïeb and Christine Léauté-Labrère
Pediatrics 2009;124:e423-e431; originally published online Aug 10, 2009;
DOI: 10.1542/peds.2008-3458









Ernemann U, et al. Current concepts in the classification, diagnosis and treatment of vascular anomalies. *Eur J Radiol* 2010; 75: 2-11.

Clasificación de las Anomalías Vasculares

Malformaciones Vasculares (I)

- **Simples**

Capilares

Venosas

Arteriales

Linfáticas

- **Combinadas**

Sme. Klippel Trenaunay

Sme. Parkes-Weber

Sme. Maffucci

Clasificación de las Anomalías Vasculares

Malformaciones Vasculares (II)

Clasificación Hemodinámica

(Jackson IT, et al. *)

Alto Flujo

Malformación Arterio-venosa

Fístula Arterio-venosa

Bajo Flujo

Malformación Capilaro-Venosa

Malformación Venosa

Malformación Linfática

*Plast Reconstr Surg 1993; 91: 1216-30. Hemangiomas, vascular malformations and lymphovenous malformations: classification and methods of treatment.

Malformación Venosa

Clínica

- Presentes al nacer
- Masa azulada, fría, compresible
- No pulsátil
- Ocasionalmente piel normal
- Asintomática / dolorosa
- Dolor: tromboflebitis
 - localización muscular
 - localización articular
- Aumento repentino: trauma
 - hemorragia
 - cambios hormonales
- Localización:

cabeza y cuello	40%
extremidades	40%
tronco	20%



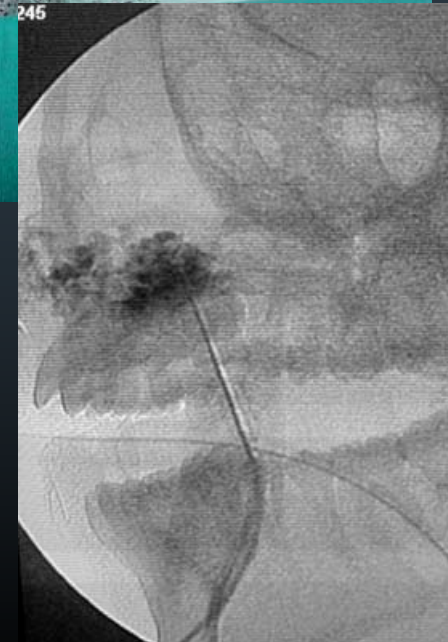
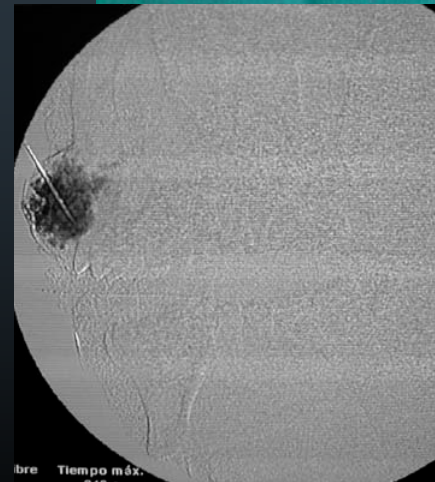
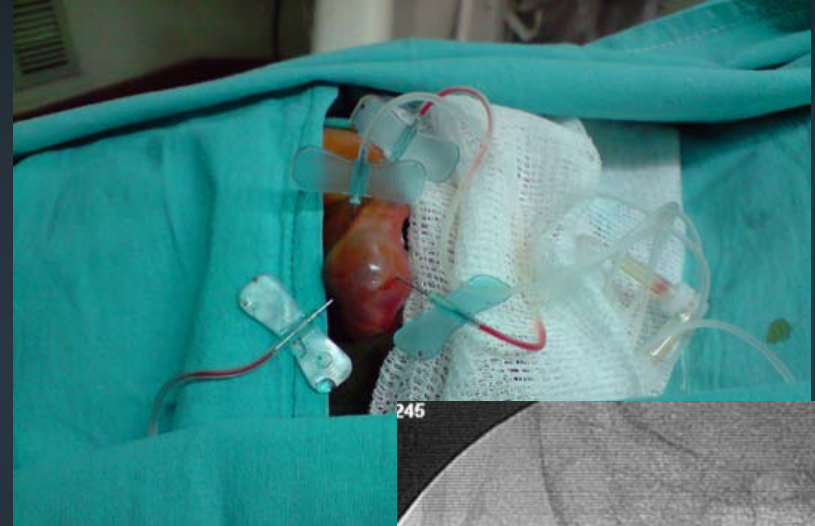
Malformaciones Venosas

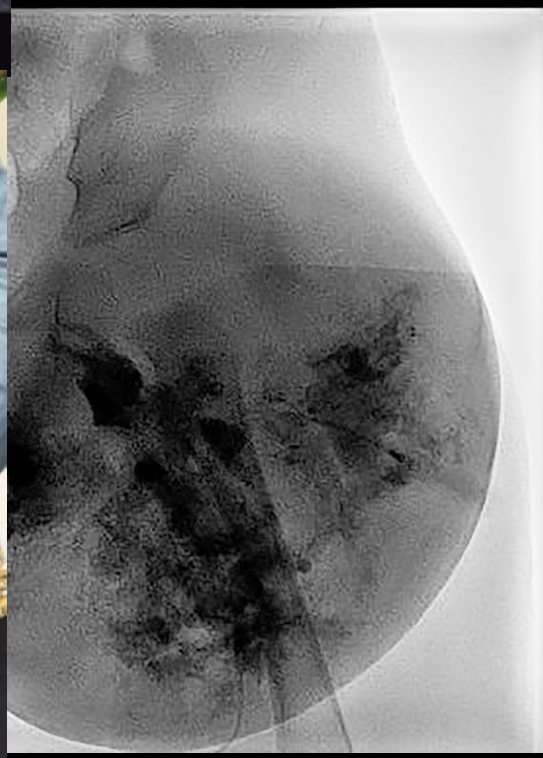
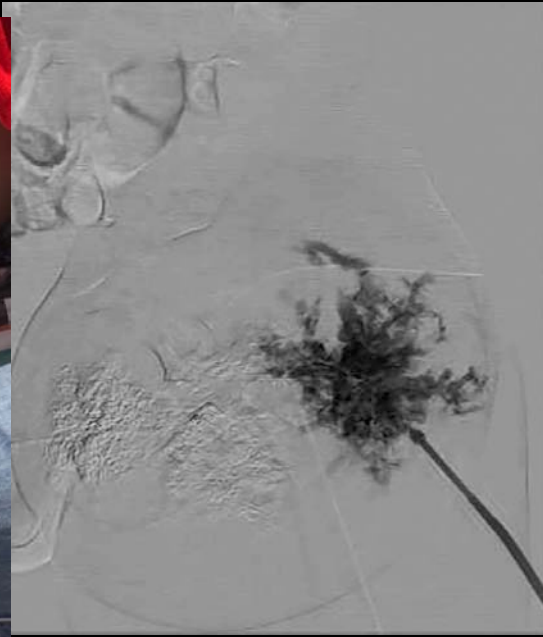
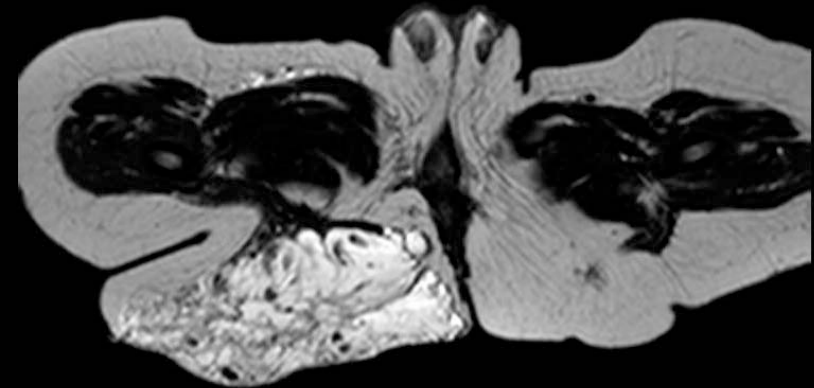
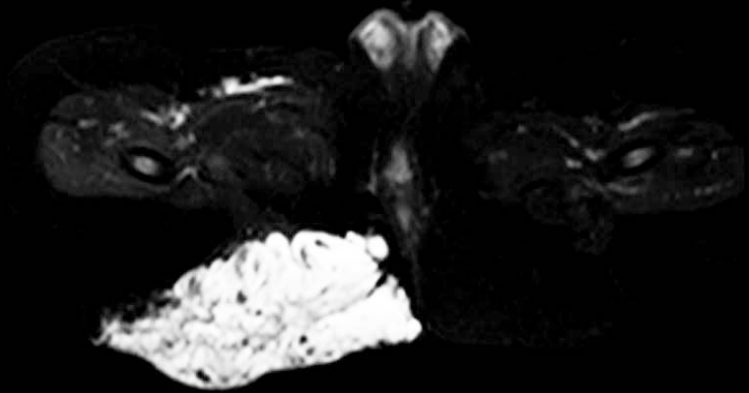
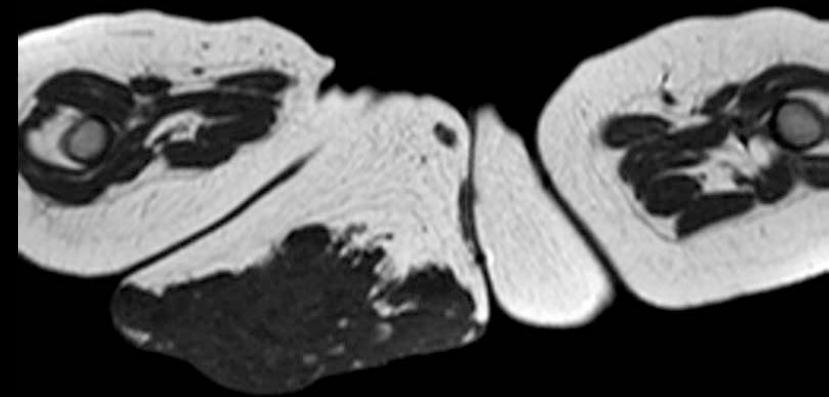
Tratamiento

- No tratamiento médico
- Cirugía: >50% recurrencia
- Trat. percutáneo (Escleroterapia)

Etanol
STS 0.5, 1, 3%
Bleomicina

- Tratamiento combinado (cirugía post-esclerosis)





Malformaciones Venosas

STUDY

Coagulation Disorders in Patients With Venous Malformation of the Limbs and Trunk

A Case Series of 118 Patients

Elisabeth Mazoyer, MD; Odile Enjolras, MD; Annouk Bisdorff, MD; Jérôme Perdu, MD; Michel Wassef, MD; Ludovic Drouet, MD

Objective: To investigate the clinical characteristics of venous malformation of the limbs and trunk and known but poorly appraised associated coagulation disorders. Venous malformations are ubiquitous, slow-flow vascular anomalies known to be occasionally painful because of thrombotic episodes inside the lesion.

Design: Large case series, with screening of accepted standard coagulation tests.

STUDY

Association of Localized Intravascular Coagulopathy With Venous Malformations

Anne Dompmartin, MD; Aurélie Acher, MD; Pascal Thibon, MD; Sébastien Tourbach, MD; Cédric Hermans, MD, PhD; Véronique Deneys, MD; Ben Pockock, MD; Agnès Lequerrec, MPhSc; Daniel Labbé, MD; Marie-Thérèse Barrellier, MD; Romain Vanwijck, MD, PhD, FRCS; Miikka Vikkula, MD, PhD; Laurence M. Boon, MD, PhD

...e which venous malformations
...gulopathy. Venous malforma-
...ular malformations present at
...avascular coagulopathy (LIC)
...osis within a lesion and severe
...cedures.

...venience sample accrued from

...pable phleboliths, and truncal localization were associ-
...ated with high D-dimer levels. In the multivariate analysis,
...only large surface area and presence of phleboliths re-
...mained independently associated with high D-dimer lev-
...els. Severe LIC, characterized by concomitant low fi-
...brinogen level, was associated with extensive venous
...malformations of the extremities.



Pediatr Clin N Am 55 (2008) 339-355

PEDIATRIC CLINICS
OF NORTH AMERICA

The Role of the Hematologist/Oncologist in the Care of Patients with Vascular Anomalies

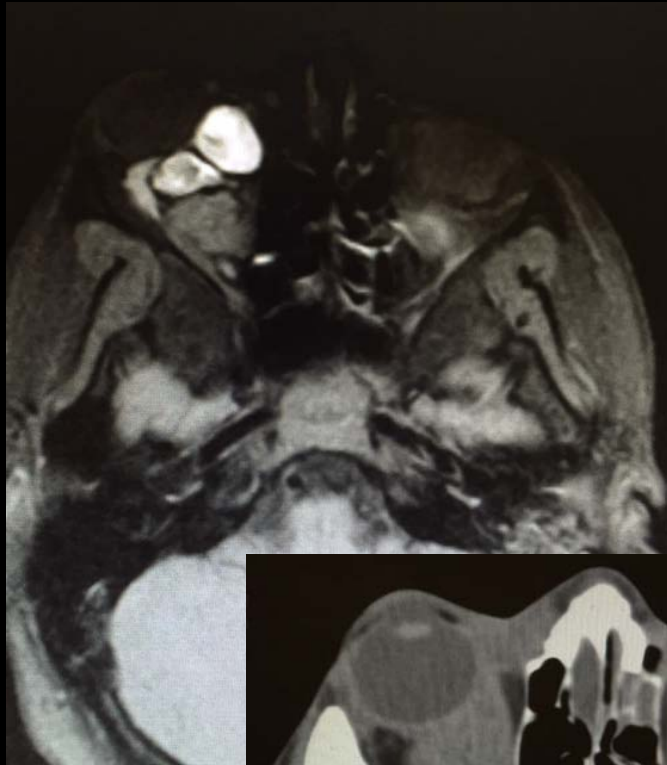
Denise M. Adams, MD^{a,b,*}, Mary Sue Wentzel, RN^b

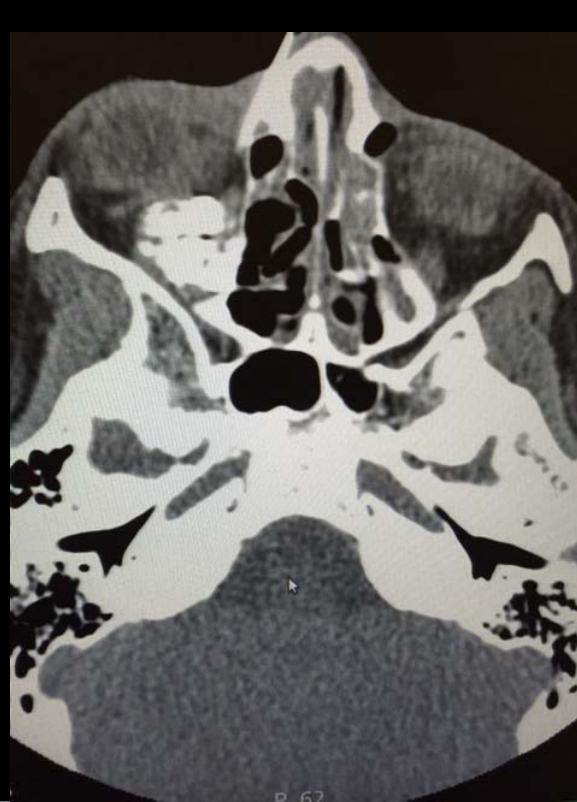
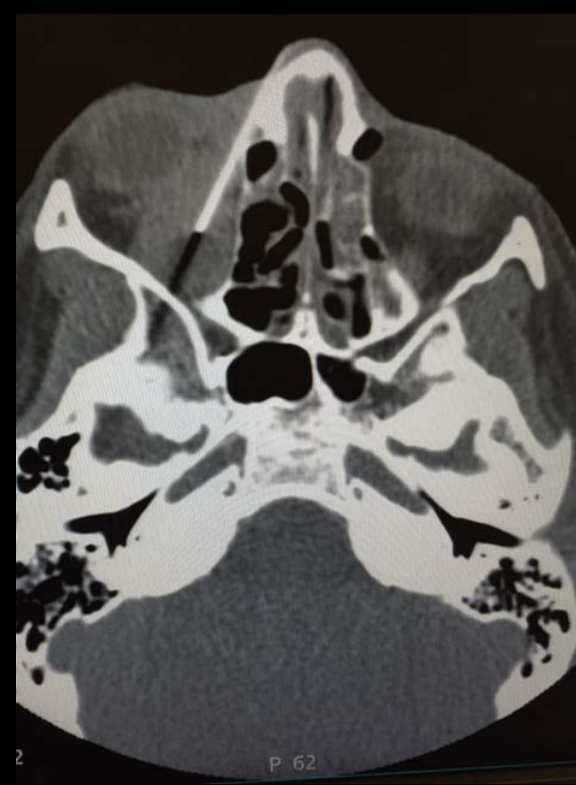
^aDivision of Hematology/Oncology, Cincinnati Children's Hospital Medical Center, University of Cincinnati, MLC 7015, 3333 Burnet Avenue, Cincinnati, OH 45229, USA

^bHemangioma and Vascular Malformation Center, Cincinnati Children's Hospital Medical Center, 3333 Burnet Avenue, Cincinnati, OH 45229, USA

Arch Dermatol 2008; 144: 861-867

Arch Dermatol 2008; 144: 873-877

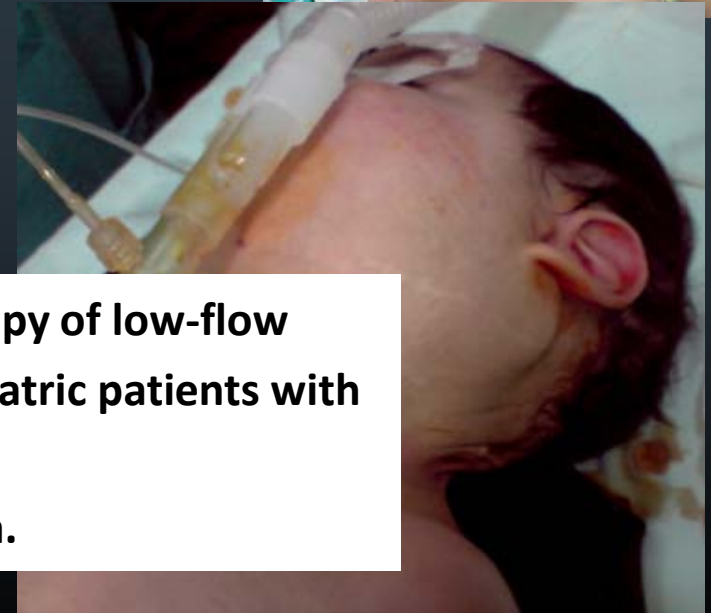




Malformaciones Vasculares Cervicales y Faciales

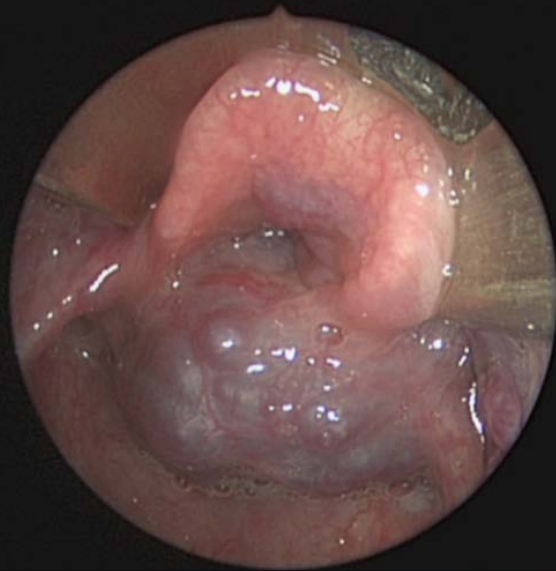
Traqueostomía

- Compromiso de la vía aérea
- MV extensas que rodean la tráquea
- Previo al trat. esclerosante

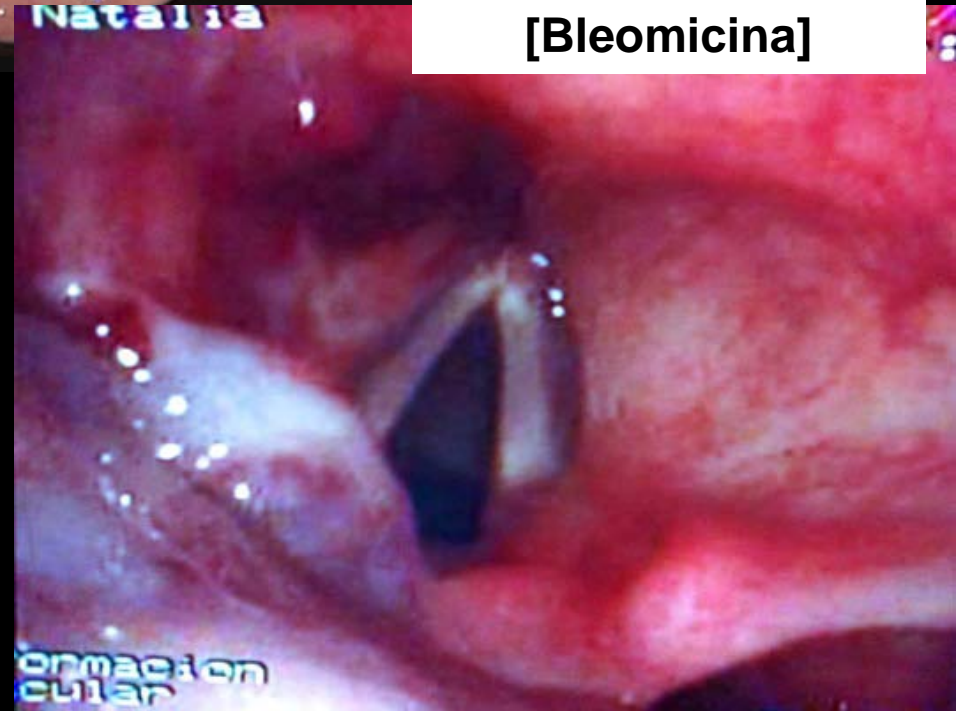
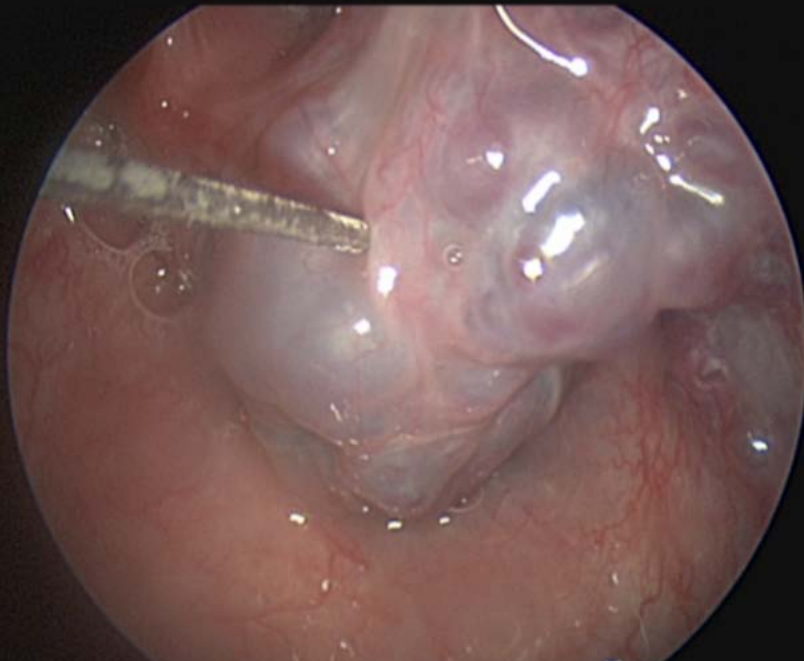


"Avoiding Tracheostomy": Percutaneous sclerotherapy of low-flow vascular malformations involving the airway in pediatric patients with elective prolonged endotracheal intubation.

Sierre S, et al. SIR Meeting 2015, Vancouver, Canada.



**Control
2 sesiones
[Bleomicina]**



Sierre S, et al. Endoscopy-assisted percutaneous treatment of laryngeal low flow vascular malformations. J Vasc Interv Radiol 2014; 25: S129.

Malformación linfática

Clínica

- Masa de partes blandas
- Gralmente, aparición antes 2 años
- Piel: normal / vesículas
- Localización
 - cabeza y cuello 70%
 - región axilar 20%
 - extremidades/retroperitoneo/pelvis
- Aumento de tamaño
 - infección
 - sangrado
- Tipos
 - Macroquístico
 - Microquístico
 - Mixto



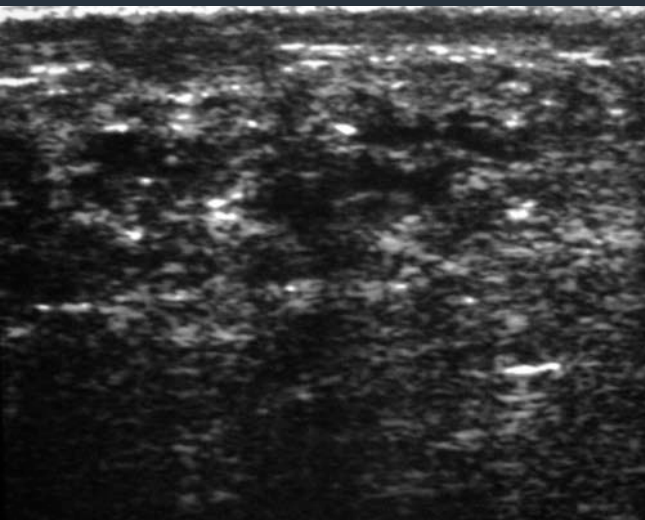
Malformación linfática

Ecografía

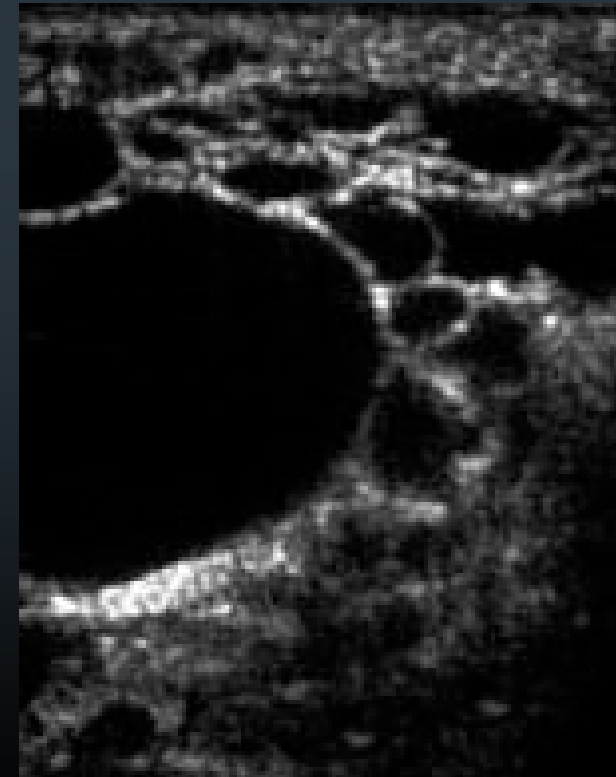


- **Macroquístico**
(grandes quistes de diferentes tamaños)

- **Microquístico**
(lesión hiperecogénica)



- **Mixto**

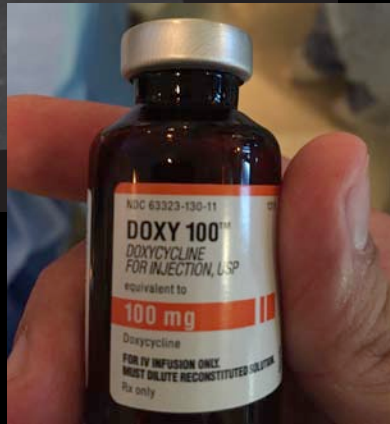
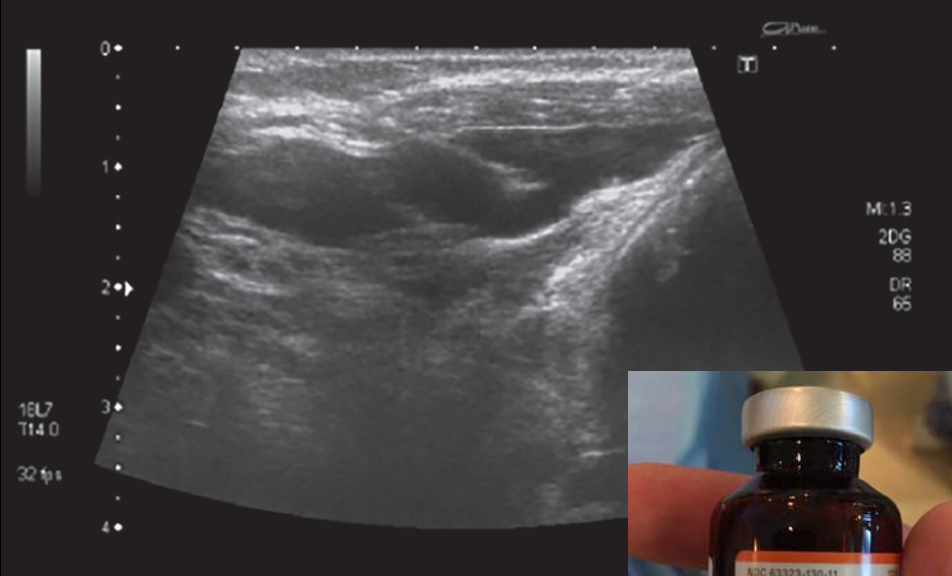
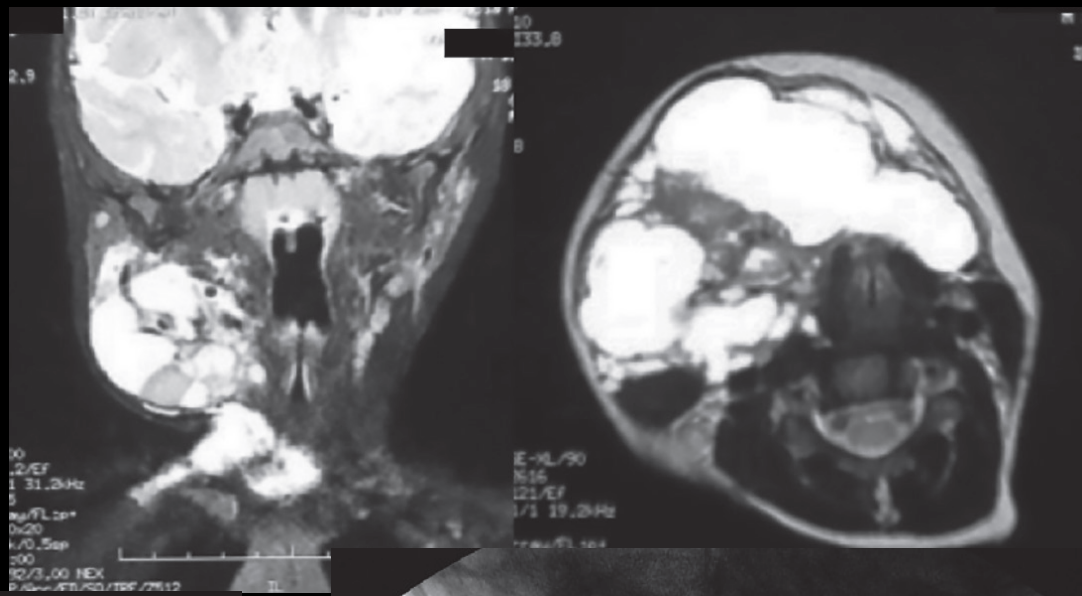


Malformación linfática

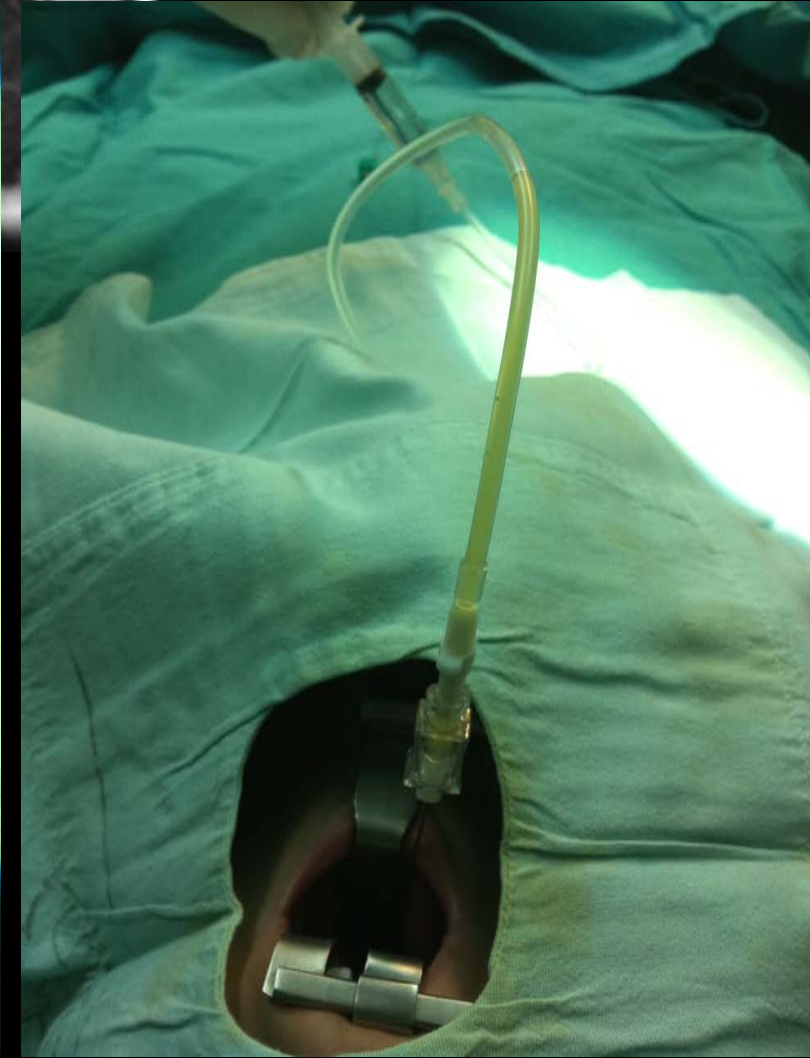
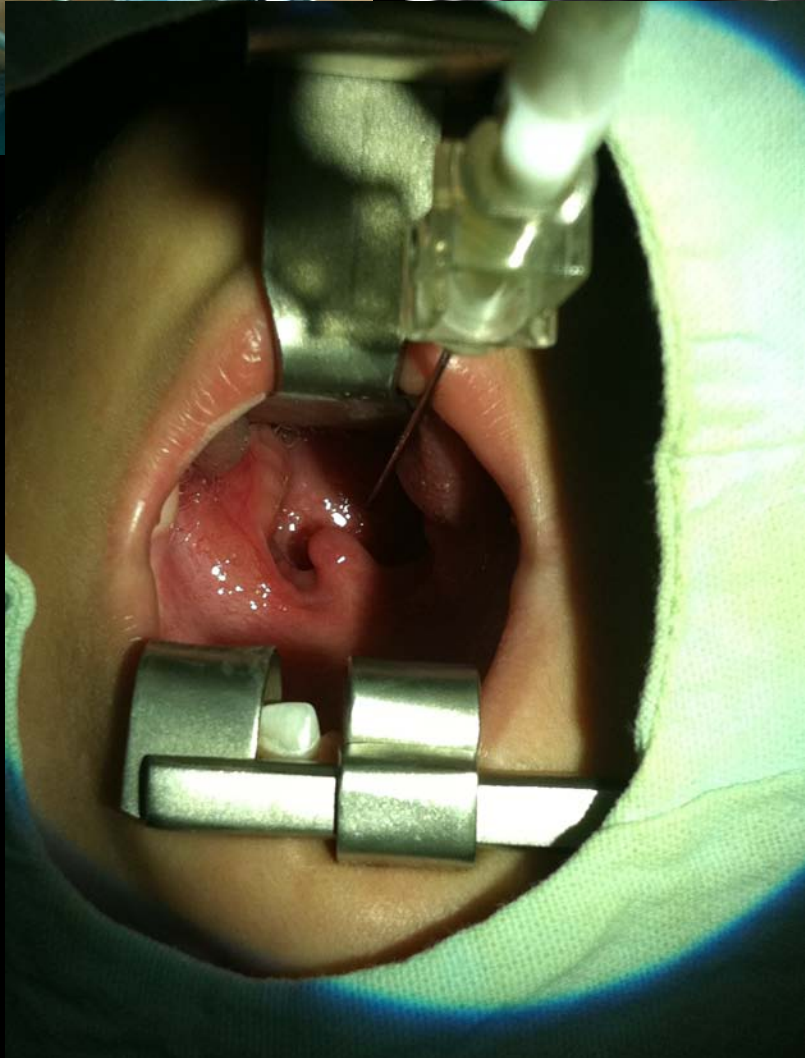
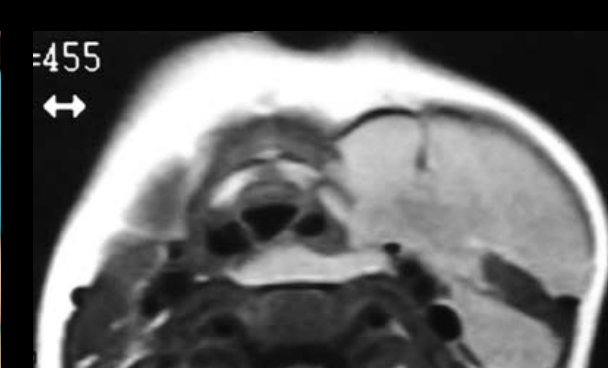
Tratamiento

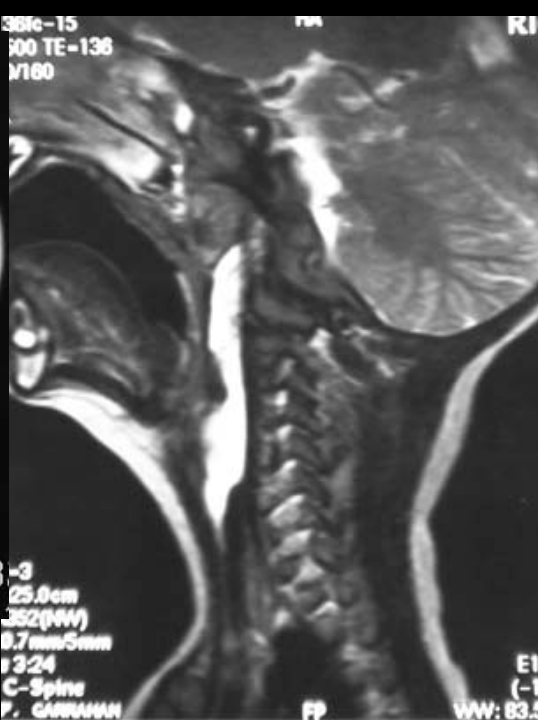
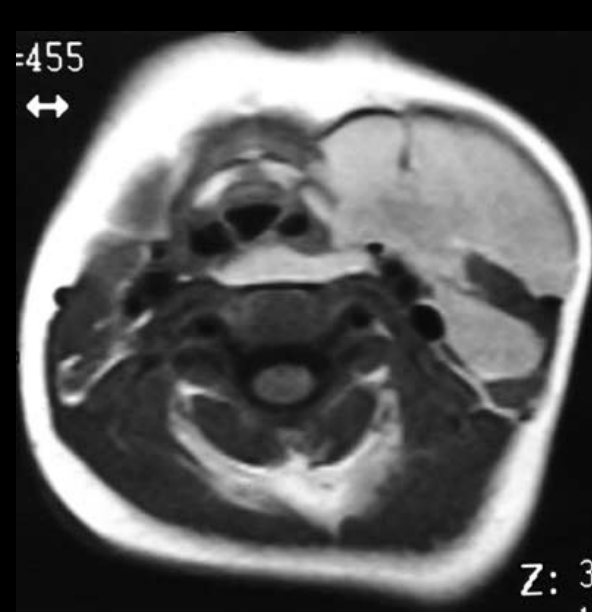
- **Macroquístico**
Esclerosis percutánea
(doxiciclina, STS, bleomicina, etanol)
- **Microquístico**
esclerosis / cirugía
(bleomicina)
- **Mixtas**
esclerosis / cirugía
(STS, doxiciclina, bleomicina)



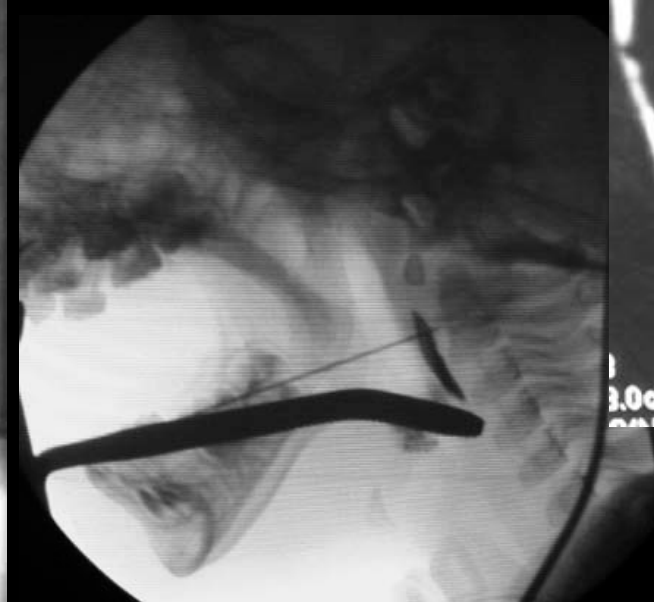








Z: 3-3
25.0cm
1.352(NW)
0.7mm/5mm
1324
C-Spine
P. GUNAWAN





4 (09:26)
17-1



6 (09:32)
21



4 (09:26)
25-1



4 (09:26)
34-1



30 (10:44)
47-1



30 (10:44)
38-1



Sildenafil for Severe Lymphatic Malformations

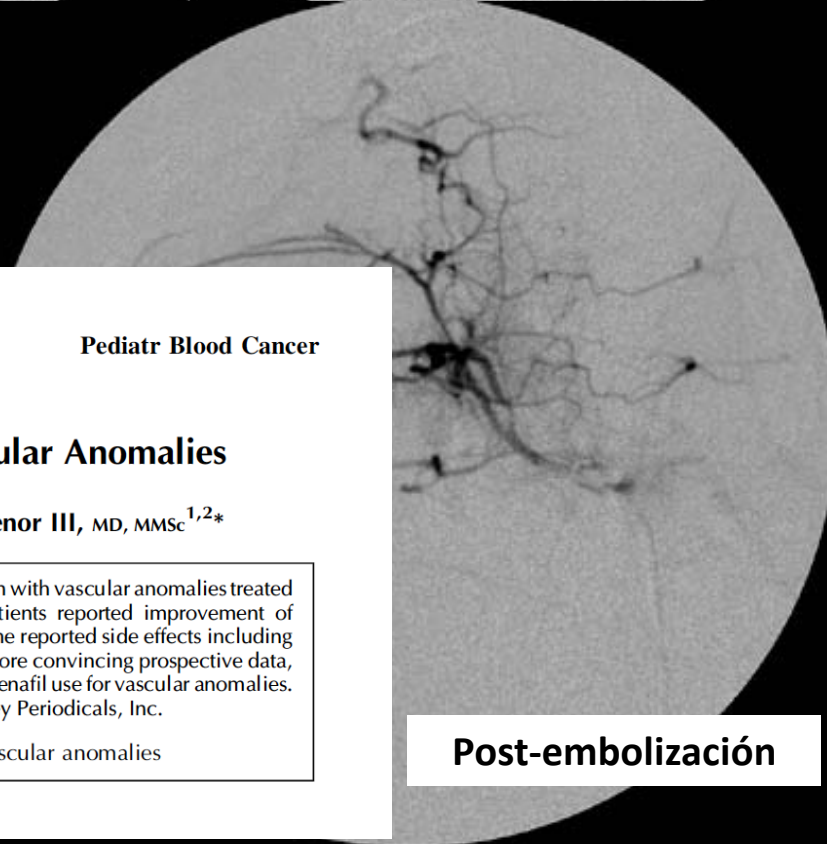
TO THE EDITOR: Lymphatic malformations are uncommon congenital vascular anomalies that can cause complications including obstruction of vital organs and their function, recurrent infection, and disfigurement.¹ Current procedural treatments are only partially successful, and lymphatic

presence of only minimal residual lymphatic malformation (Fig. 1B).

Figure 1 (facing page). MRI of the Chest, Performed

Swetman GL, Berk DR. N Eng J Med 2012; 366: 4

Photographs of Subject 2 before and after Treatment



Pediatr Blood Cancer

BRIEF REPORT

Caution Is Recommended Prior to Sildenafil Use in Vascular Anomalies

Hannah Rankin, BS,^{1,2} Kelley Zwicker, MSc, MD, FRCPC,^{1,2,3} and Cameron C. Trenor III, MD, MMSc^{1,2*}

Since publication of a single case report of lymphatic malformation improvement during sildenafil therapy for pulmonary hypertension, sildenafil use has propagated across multiple vascular anomalies diagnoses. Vascular anomalies are rare conditions, often with poor long-term outcomes from available therapies, making these patients vulnerable to novel therapy use. We have

retrospectively reviewed 14 children with vascular anomalies treated with sildenafil. None of these patients reported improvement of disease while on treatment and some reported side effects including infections and bleeding. Pending more convincing prospective data, we recommend caution prior to sildenafil use for vascular anomalies. Pediatr Blood Cancer © 2015 Wiley Periodicals, Inc.

Key words: lymphatic malformation; medical therapy; natural history; sildenafil; vascular anomalies

Post-embolización

2 (12:27)
19-1

10 (12:46)
15-1

Sirolimus for the Treatment of Complicated Vascular Anomalies in Children

Adrienne M. Hammill, MD, PhD,^{1,2*} MarySue Wentzel, RN,¹ Anita Gupta, MD,^{1,3} Stephen Nelson, MD,⁴ Anne Lucky, MD,^{1,5} Ravi Elluru, MD, PhD,^{1,6} Roshni Dasgupta, MD,^{1,7} Richard G. Azizkhan, MD,^{1,7} and Denise M. Adams, MD^{1,2}

Background. Vascular anomalies comprise a diverse group of diagnoses. While infantile hemangiomas are common, the majority of these conditions are quite rare and have not been widely studied. Some of these lesions, though benign, can impair vital structures, be deforming, or even become life-threatening. Vascular tumors such as kaposiform hemangioendotheliomas (KHE) and complicated vascular malformations have proven particularly difficult to treat. **Procedure.** Here we retrospectively evaluate a series of six patients with complicated, life-threatening vascular anomalies who were

treated with the mTOR inhibitor sirolimus for compassionate use at two centers after failing multiple other therapies. **Results.** These patients showed significant improvement in clinical status with tolerable side effects. **Conclusions.** Sirolimus appears to be effective and safe in patients with life-threatening vascular anomalies and represents an important tool in treating these diseases. These findings are currently being further evaluated in a Phase II safety and efficacy trial. *Pediatr Blood Cancer* 2011;57:1018–1024. © 2011 Wiley-Liss, Inc.

Key words: vascular a

Pediatr Blood Cancer 2011;57:904–905

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lar Anomalies

rapy for Pediatric Vascular

Sirolimus for the Treatment of a Massive Capillary-Lymphatico-Venous Malformation: A Case Report

Aleksandar M. Vlahovic, Natasa S. Vlahovic and Emir Q. Haxhija
Pediatrics 2015;136:e513; originally published online July 6, 2015;
DOI: 10.1542/peds.2014-3469

Heather Mills Soni, CPNP¹ Sheena Pimpalwar, MD²

as Children's Cancer and Hematology
logy, Department of Radiology, TCH,
uston, Texas

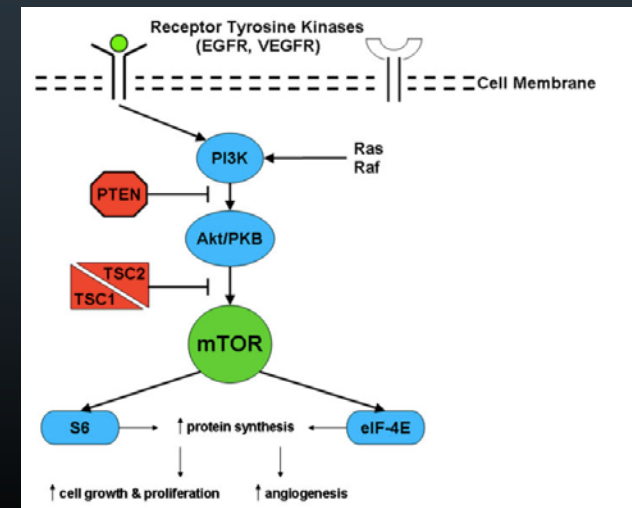
Address for correspondence: Judith F. Margolin, MD, Department of Pediatrics, Baylor College of Medicine, Texas Children's Cancer and Hematology Service, Suite 1510, Clinical Care Center, 6701 Fannin Street, Houston, Texas 77030 (e-mail: jfmargol@txch.org).

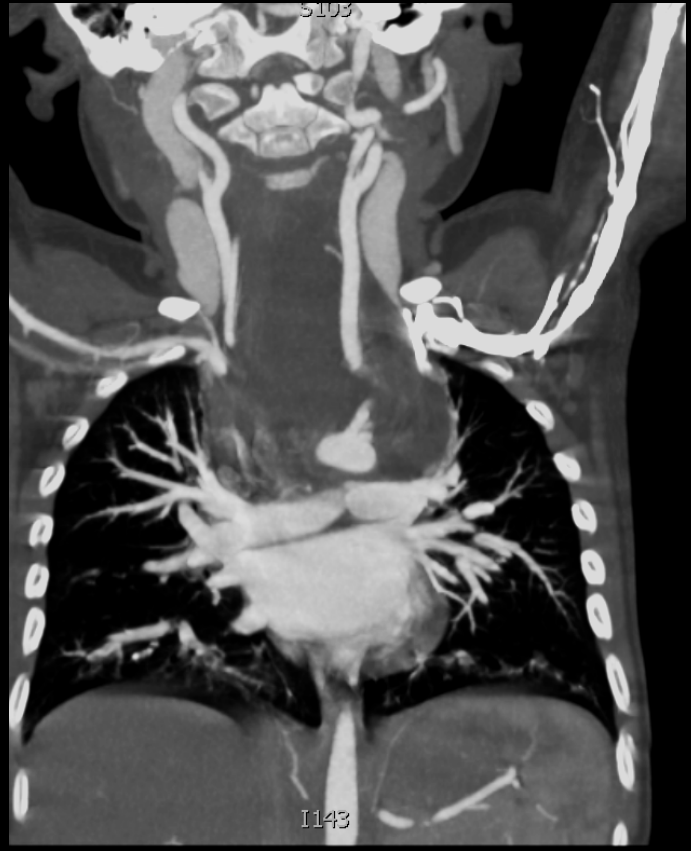
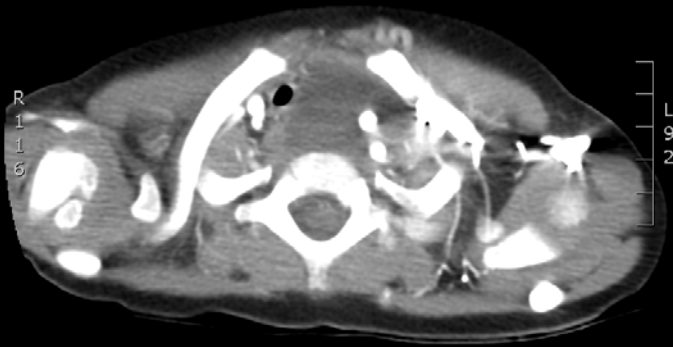
Sirolimus / Rapamicina

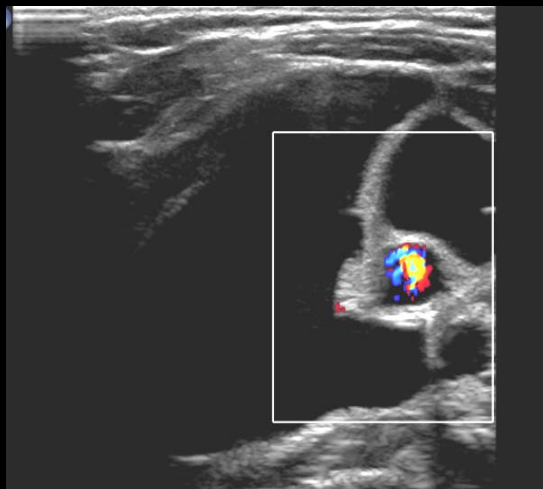
- **Medicamento inmunosupresor**
trasplante renal
- **Eficacia para freno de la proliferación celular y el**
crecimiento tumoral
cubierta de stents p/ disminuir restenosis

Sirolimus: Mecanismo de Acción

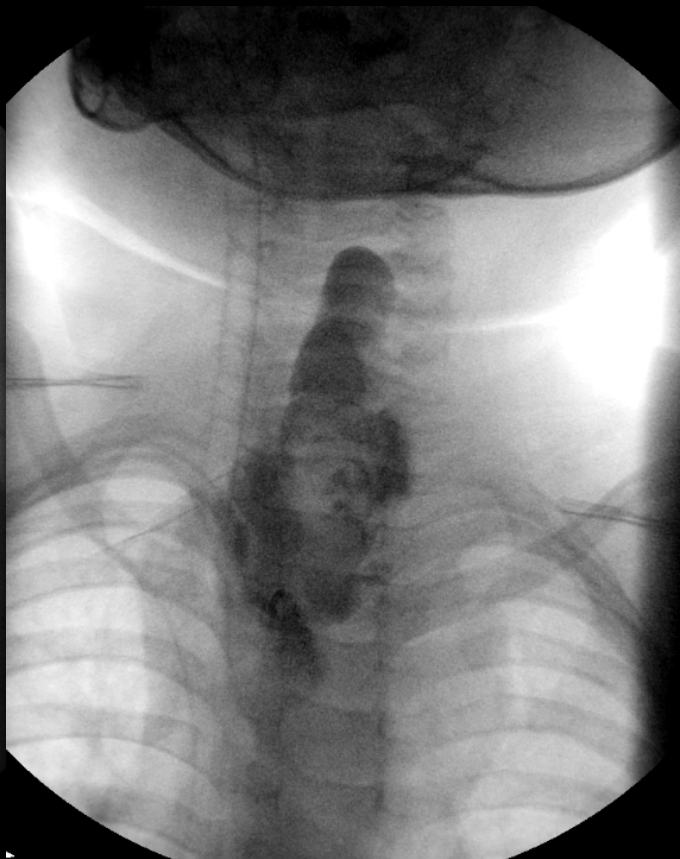
- mTOR es una proteína que regula el crecimiento, la proliferación, la motilidad y la supervivencia celulares
- Participación en la síntesis de proteínas
- Básicamente, se trata de un inhibidor de la angiogénesis

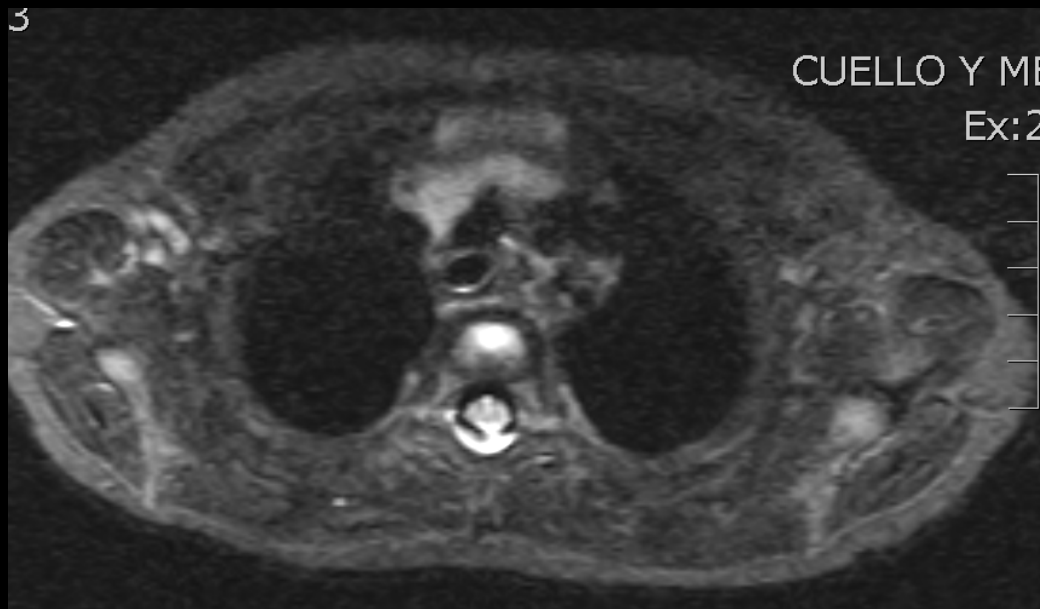


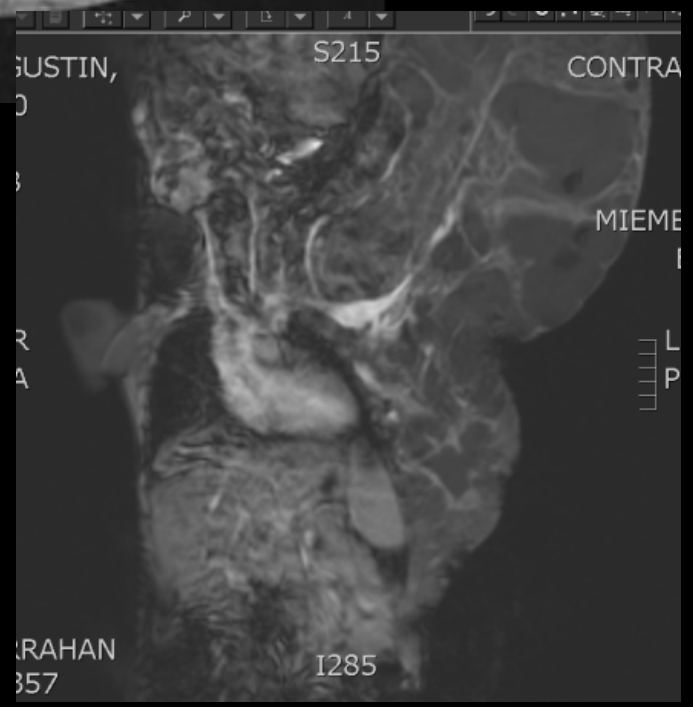
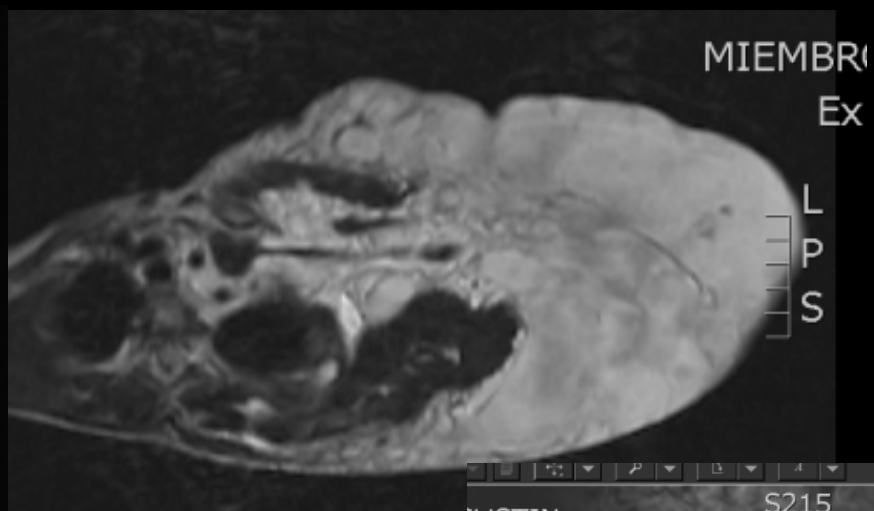
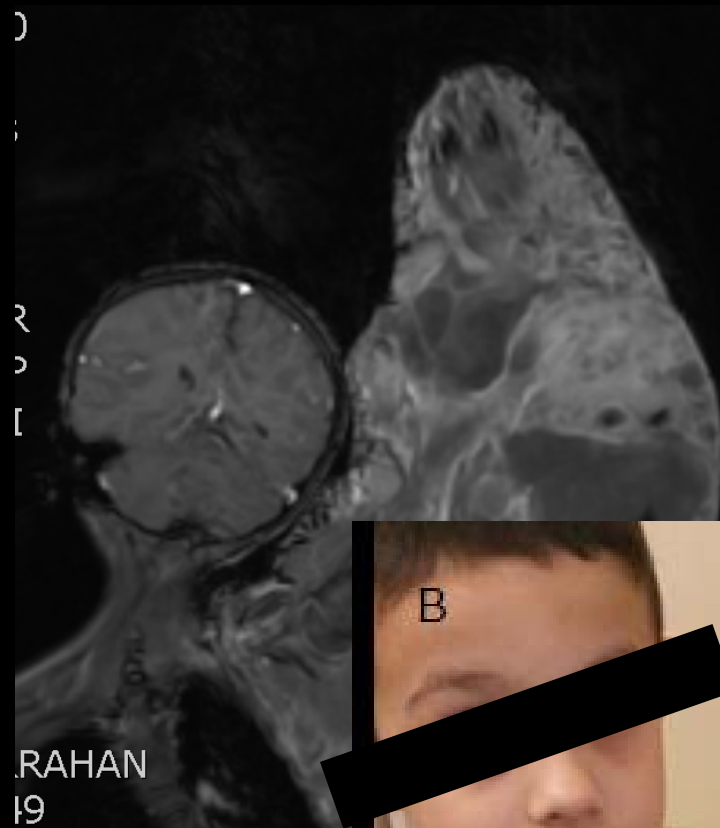




(12:30)











Pre-Sirolimus

Post-Sirolimus

Malformaciones Vasculares

Consideraciones finales

- **Patología rara, pero importante**
- **Una clasificación (Mulliken y Glowacki)**
- **Imágenes juegan un papel esencial**
- **Diagnóstico correcto y preciso es indispensable par un tratamiento efectivo**
- **Equipos multidisciplinarios**

Malformaciones Vasculares

Consideraciones finales

- **Rx Intervencionista constituye un pilar del tratamiento**
- **Planificación / traqueostomía / intubación prolongada**
- **Actualización constante**
- **Manejo multidisciplinario**