

# VIII SISAV International Congress



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Vascular anomalies:  
update on diagnostic  
and therapeutic approach

Roma

9-11 / 9 / 2021

Auditorium Valerio Nobili  
Ospedale Pediatrico Bambino Gesù  
(Sede di S. Paolo)  
Viale Ferdinando Baldelli, 38

Immagine per gentile concessione della Fabbrica di San Pietro in Vaticano



# EMANGIOMA INFANTILE LARINGEO

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# Laryngeal Hemangioma

**60% of infantile hemangiomas (IH) are located in the head and neck regions**

+/- cutaneous IH, with a particular distribution in 50% of cases (“beard” area or S3 area - chin, anterior neck and lower lip)

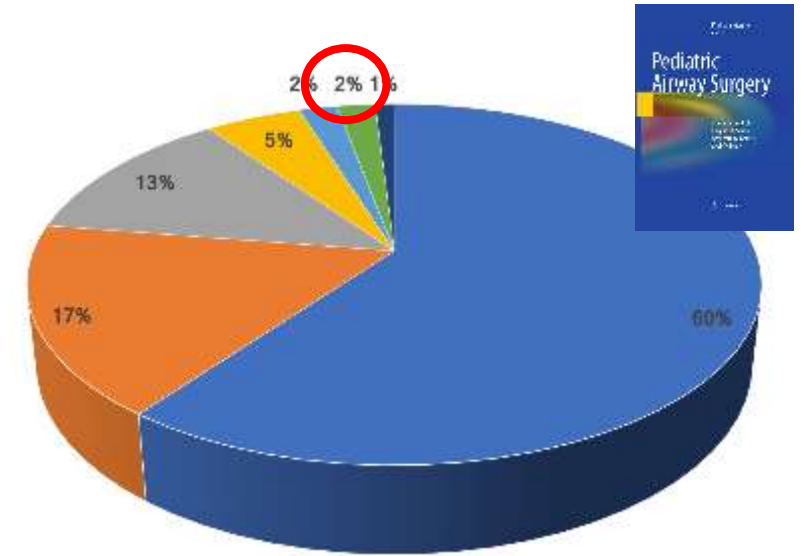
Haggstrom and Frieden’s classification

Haggstrom A.N. et al. Infantile hemangiomas: New clues to hemangioma pathogenesis and embryonic facial development. *Pediatrics* 117, 698–703 (2006).

## Laryngeal Hemangioma

### > Sub-Glottic Hemangiomas (SGH)

Rare benign tumour of the airway, accounting for only **1.5-2%** of all congenital laryngeal anomalies



- Laryngomalacia
- Vocal Cord Paralysis
- Subglottic Stenosis
- Web
- Hemangioma
- Saccular Cyst/Laryngoceles
- Cleft



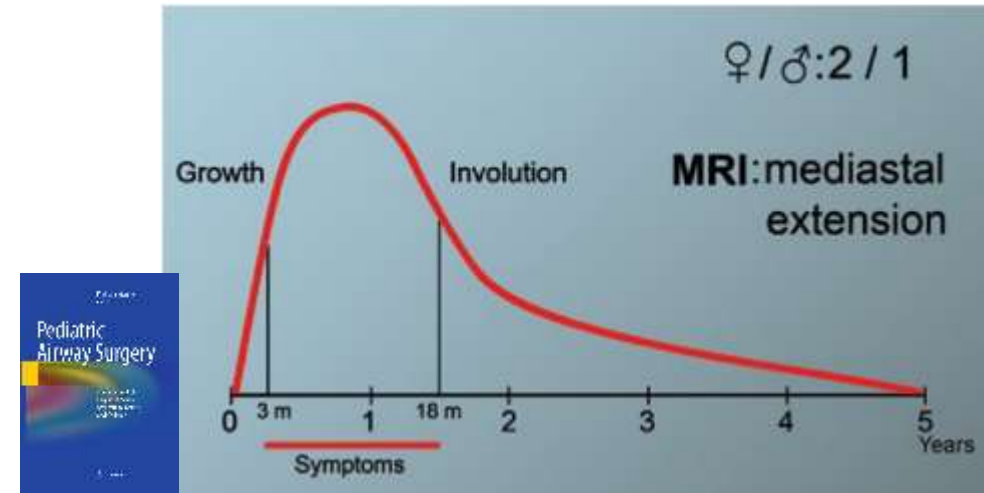
- **BIPHASIC AND WORSENING STRIDOR**
- Hoarseness
- Barking cough
- Tirage
- Chest retractions
- Failure to thrive
- Sleep apnea
- Respiratory failure

**THE AGE OF ONSET**  
**IS TYPICAL - 2-4 months**

O-Lee, T. J. & Messner, A. Subglottic Hemangioma. *Otolaryngologic Clinics of North America* vol. 41 903–911 (2008)

- ✓ Rapid growth
- ✓ Age 10-12 months - Plateau of the symptoms progression
- ✓ Then the symptoms decrease slowly
- ✓ The symptoms disappear around the age of 2 years
- ✓ 5-10 years complete resolution of the tumour

Bruckner, A.L., Frieden, I.J.: *Infantile hemangiomas*. *J. Am. Acad. Dermatol.* 55, 671–682 (2006)



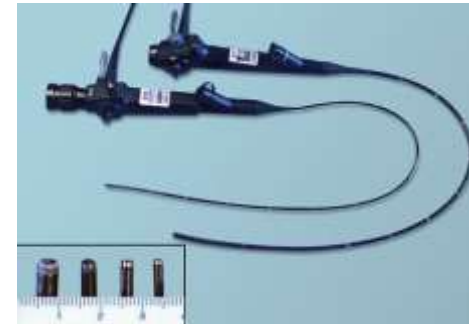


## Airway Endoscopy (diagnosis and follow-up)

### 1. AWAKE

*Upper airway*

*Laryngeal motility and respiratory space*



### 2. GENERAL ANESTHESIA

*Cricoarytenoid joint*

*Subglottis*

*Trachea*



- **Biopsy is not necessary in almost all cases**
- **The diagnosis is based on clinical and endoscopic findings**

*Bitar, M.A., Moukarbel, R.V., Zalzal, G.H.: Management of congenital subglottic hemangioma: trends and success over the past 17 years. Otolaryngol. Head Neck Surg. 132, 226– 231 (2005)*



## Airway Endoscopy (diagnosis and follow-up)

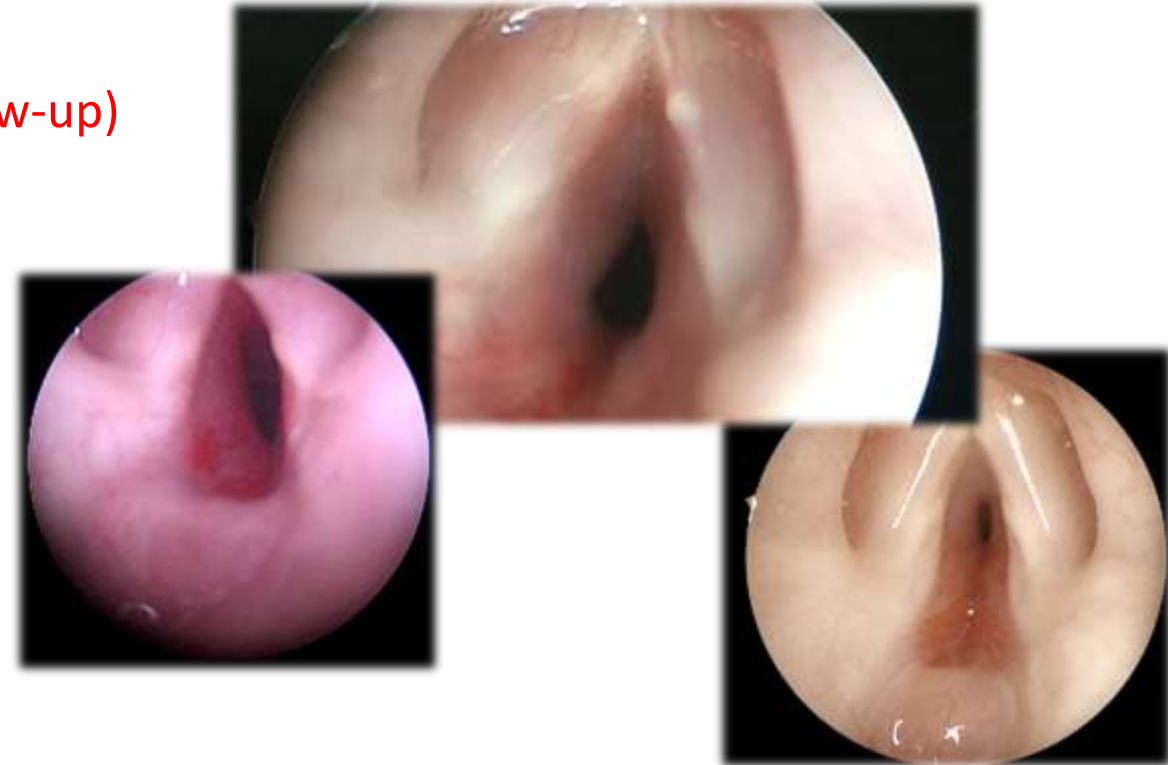




## Airway Endoscopy (diagnosis and follow-up)

The mass is:

- **Reddish and smooth**
- **Elastic/spongy and compressible**



- Airway IHs usually involve the subglottis (**the narrowest part of the pediatric airway**)
- **> located in the left posterolateral part**

*Rahbar, R. et al. The biology and management of subglottic hemangioma; past, present, future. Laryngoscope 114, 1880–1891 (2004)*





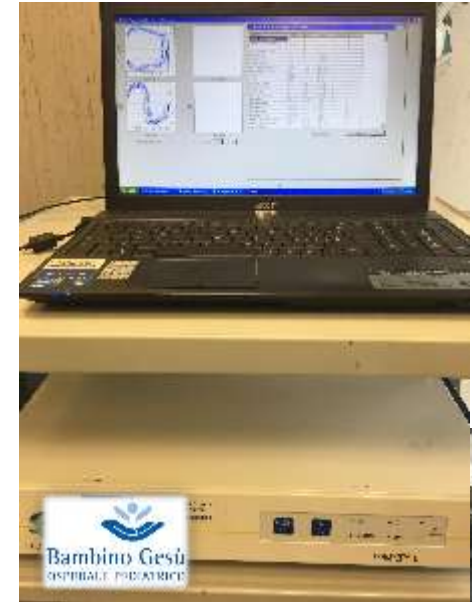
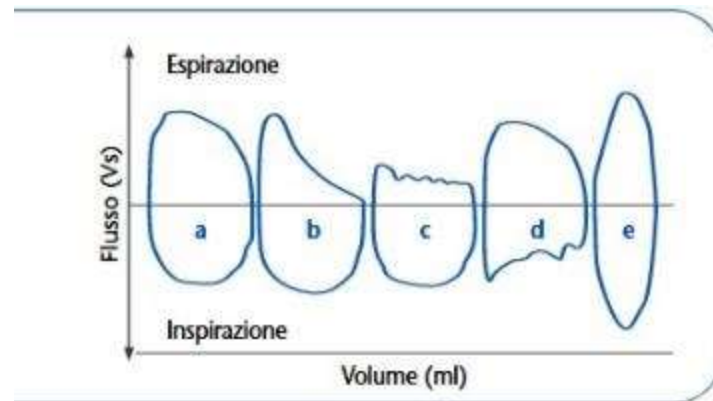
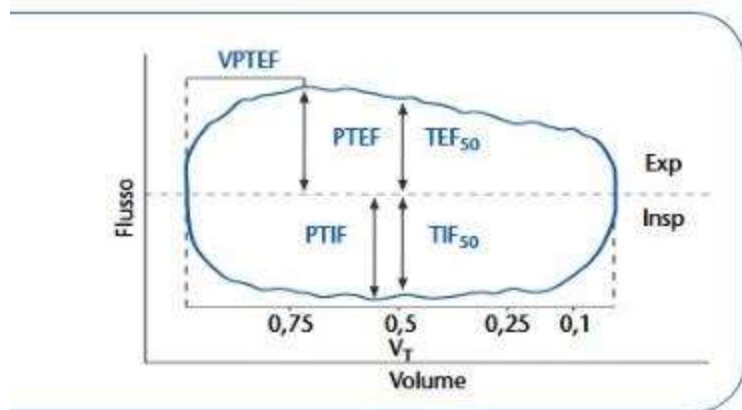
## Pulmonary Function Tests (PFTs)

< 2 years

- Spontaneous sleeping
- Facial mask
- Tidal volume and obstruction pattern/degree evaluation

> 5 years

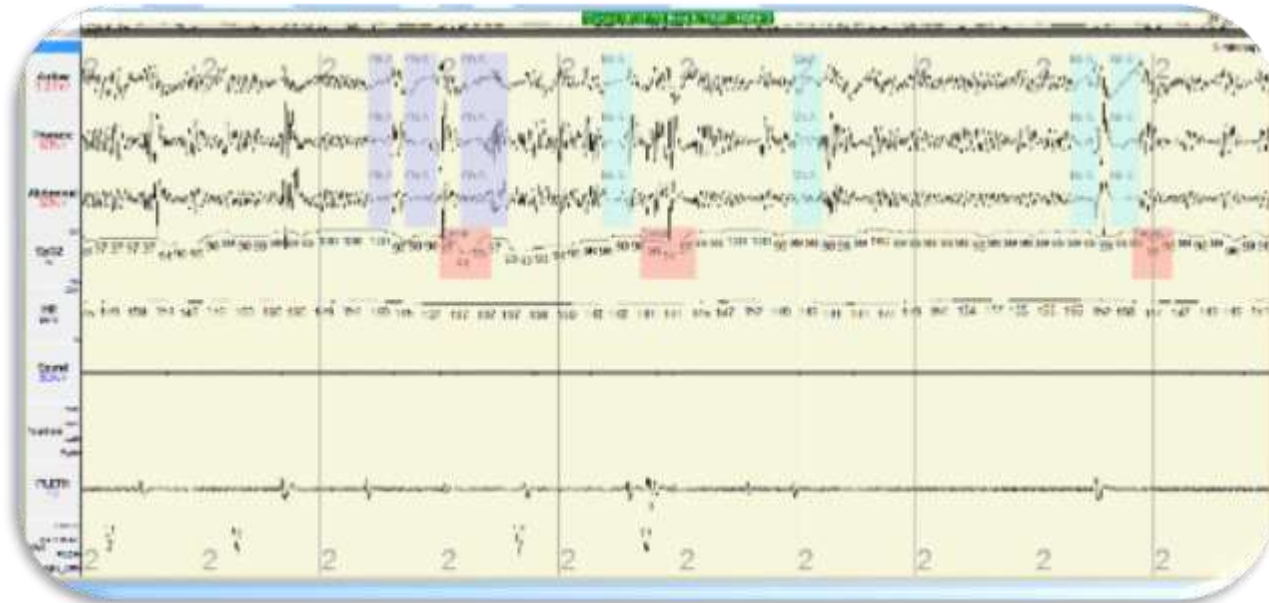
- Spirometry (basal/exercise stress test)
- Bronchodilator test
- Evaluation of flows and volumes



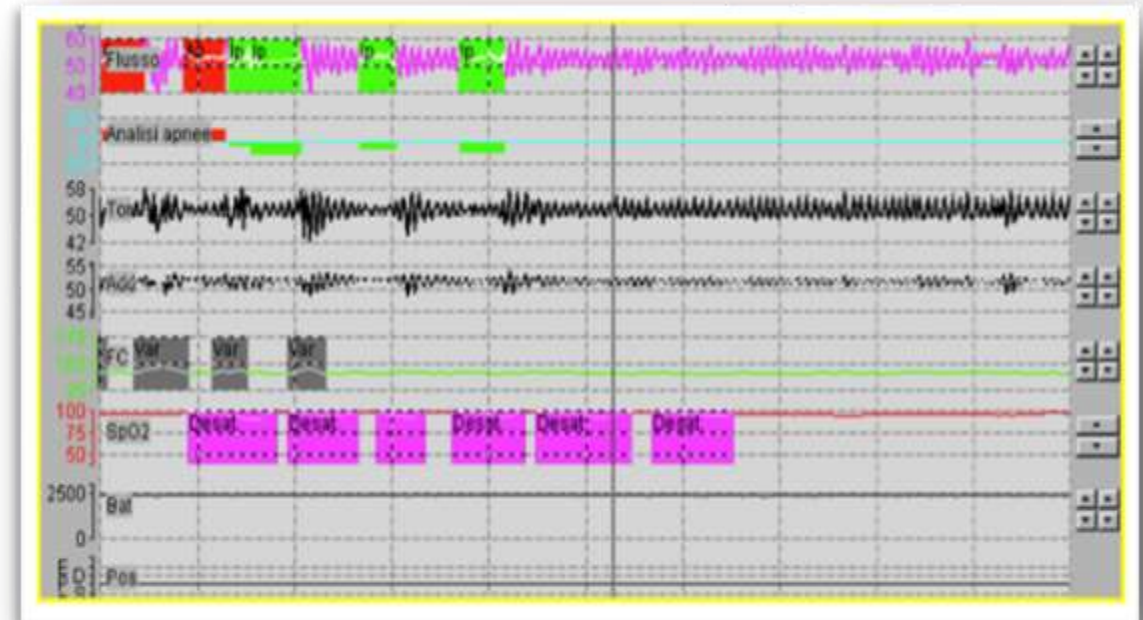


## Sleep Study Obstructive Apneas

Polysomnography



Pulse-Oximetry



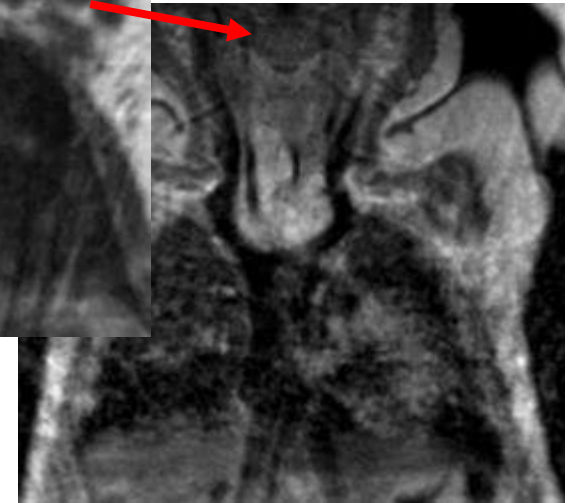
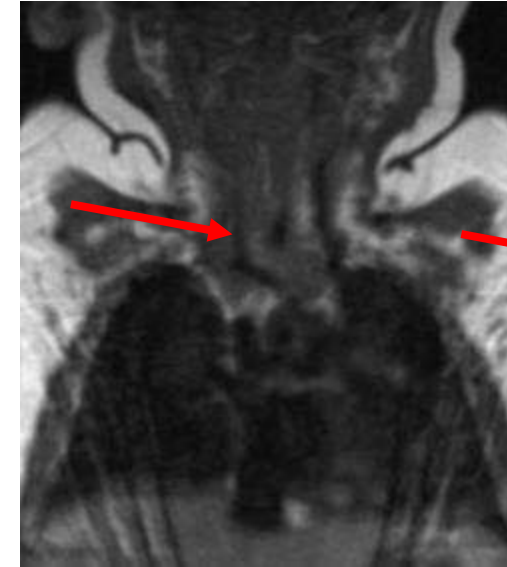
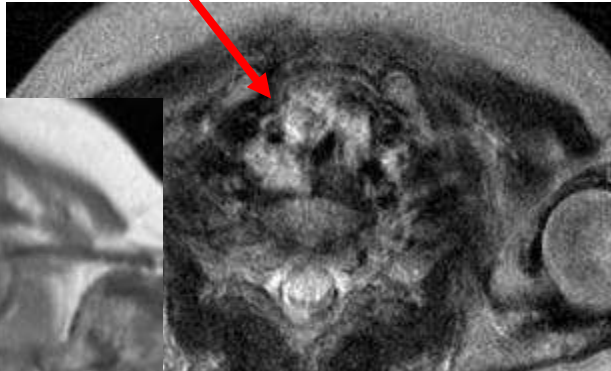
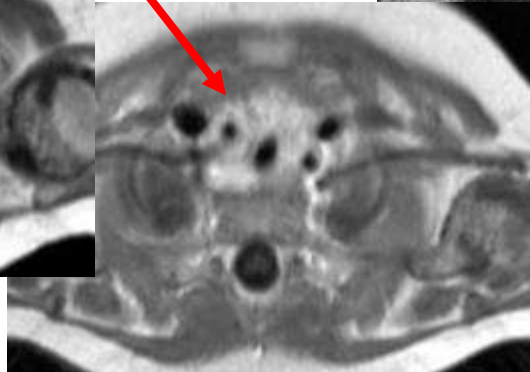
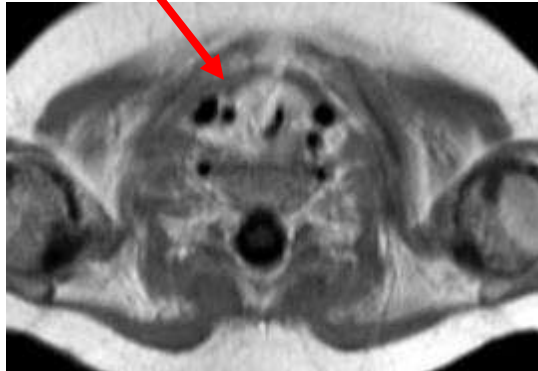




## Radiology

Contrast-enhanced MRI or CT scan for:

- therapy-resistant hemangioma
- differential diagnosis with other laryngo-tracheal diseases
- identifying potential extralaryngeal extension or extension into the upper mediastinum



T2 SEQUENCE: increase in signal intensity is typical of the proliferative phase

*Sie Stampakopoulou Otolaryngol CI North Am, Vol 33 (1) Feb 2000*



## Medical Therapy

### PROPRANOLOL (since 2008)

*Leaute-Labreze, C., de la Roque, D.: Propranolol for severe hemangiomas of infancy. N. Engl. J. Med. 358, 2649–2651 (2008)*

*Jepson, C.G., et al.: Successful treatment of isolated subglottic haemangioma with propranolol alone. Int. J. Pediatr. Otorhinolaryngol. 73, 1821–1823 (2009)*

- Non-selective  $\beta$ -blocker
- 2 mg/kg (1-3 mg/kg) of bodyweight per day
- **It radically changed the prognosis**
- Oral propranolol is now considered the gold-standard of treatment





## Medical Therapy

### SYSTEMIC STEROIDS

- <25% of effectiveness as single therapy (Replaced by Propranolol)
- It often precedes the therapy with Propranolol, especially in non-reference centers
- Stop as soon as Propranolol is started
  - ✓ Adjuvant therapy for a short period
  - ✓ Not recommend for longer than 3 weeks (long-term side effects)

Rahbar, R., Nicollas, R., Roger, G., et al.: *The biology and management of subglottic hemangioma: past, present, future. Laryngoscope* 114, 1880–1891 (2004)

Rutter, M.J.: *Laryngeal webs and subglottic hemangiomas. In: Graham, J.M., Scadding, G.K., Bull, P.D. (eds.) Pediatric ENT, pp. 211–222. Springer, Berlin/Heidelberg (2008)*



## Surgical Treatment

- Patients not responsive to treatment with propranolol
- Contraindication to  $\beta$ -blocker

## LOCAL STEROID INJECTION + INTUBATION

- High success rate in controlling the disease and respiratory complications
- Least invasive technique
- Sometimes used as adjuvant treatment to endoscopic surgery
- **BUT** often the procedure must be repeated in case of worsening



## Surgical Treatment

- Patients not responsive to treatment with propranolol
- Contraindication to  $\beta$ -blocker

## CO2 LASER RESECTION

- Rapid vaporization of the tumor
- Significant success rate (up to 95%)

*Bitar, M.A., Moukarbel, R.V., Zalzal, G.H.: Management of congenital subglottic hemangioma: trends and success over the past 17 years. Otolaryngol. Head Neck Surg. 132, 226– 231 (2005)*

*Hoeve L.J., Kuipers G.L.E., Verwoerd C.D.A. Management of infantile subglottic hemangioma: laser vaporization, submucous resection, intubation, or intralesional steroids? Int J of Ped Otorhinolaryngology 42 (1997) 179–186.*

*Saetti R, Silvestrini M, Cutrone C, Narne S. Treatment of congenital subglottic hemangiomas: our experience compared with reports in the literature. Arch Otolaryngol Head Neck Surg. 2008 Aug;134(8):848-51.*

- Lower risk of bleeding
- **BUT** risk of subglottic stenosis (especially with multiple reoperations)



## Surgical Treatment

- Patients not responsive to treatment with propranolol
- Contraindication to  $\beta$ -blocker

## SUBMUCOUS RESECTION

- Bilateral or circumferential sub-glottic hemangioma
- Fast-growing tumours
- Significant success rate

*Bitar, M.A., Moukarbel, R.V., Zalzal, G.H.: Management of congenital subglottic hemangioma: trends and success over the past 17 years. Otolaryngol. Head Neck Surg. 132, 226– 231 (2005)*

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- Invasive surgery
- **BUT** almost always reserved to cases resistant to other treatment (**the most invasive surgery**)



## Surgical Treatment

- Patients not responsive to treatment with propranolol
- Contraindication to beta blocker

## SUBMUCOUS RESECTION

- **Endoscopic microdebrider** to excise subglottic lesions (small tumour)
- Perichondrium and cartilage are preserved
- No secondary thermal mucosal damage

### Management of subglottic hemangioma

Seth M. Pransky and Cecilia Canto

Current Opinion in Otolaryngology & Head and Neck Surgery 2004,  
12:509–512



# Our Experience – 2009-20

2009-2020	n°	%
<b>Total</b>	<b>36</b>	
Male	9	25%
Female	27	75%
Respiratory symptoms	31	86,1%
Risk factors	8	22,2%
Cutaneous	23	63,9
PHACES	2	5,5

## THERAPY – PROPRANOLOL 1-3 mg/kg/die

28/36 - Propranolol 2 mg/kg/die

7/36 - Propranolol 3 mg/kg die

1/36 - Propranolol 1 mg/kg/die

10/36 - Additional therapy with **steroids**

5/36 (13.9%) had **relapses at suspension** and restarted oral therapy

**NO** propranolol non-responder in our cohort





# Our Experience – 2009-20

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DURATION OF THERAPY: 6-26 months (average 15 m)

## AGE OF:

- Onset of symptoms - 0-90 days (average 30,9 d)
- Start of therapy - 1-10 months (average 3,5 m)
- Suspension of therapy - 10-54 months (average 20 m)



# Our Experience – 2009-20

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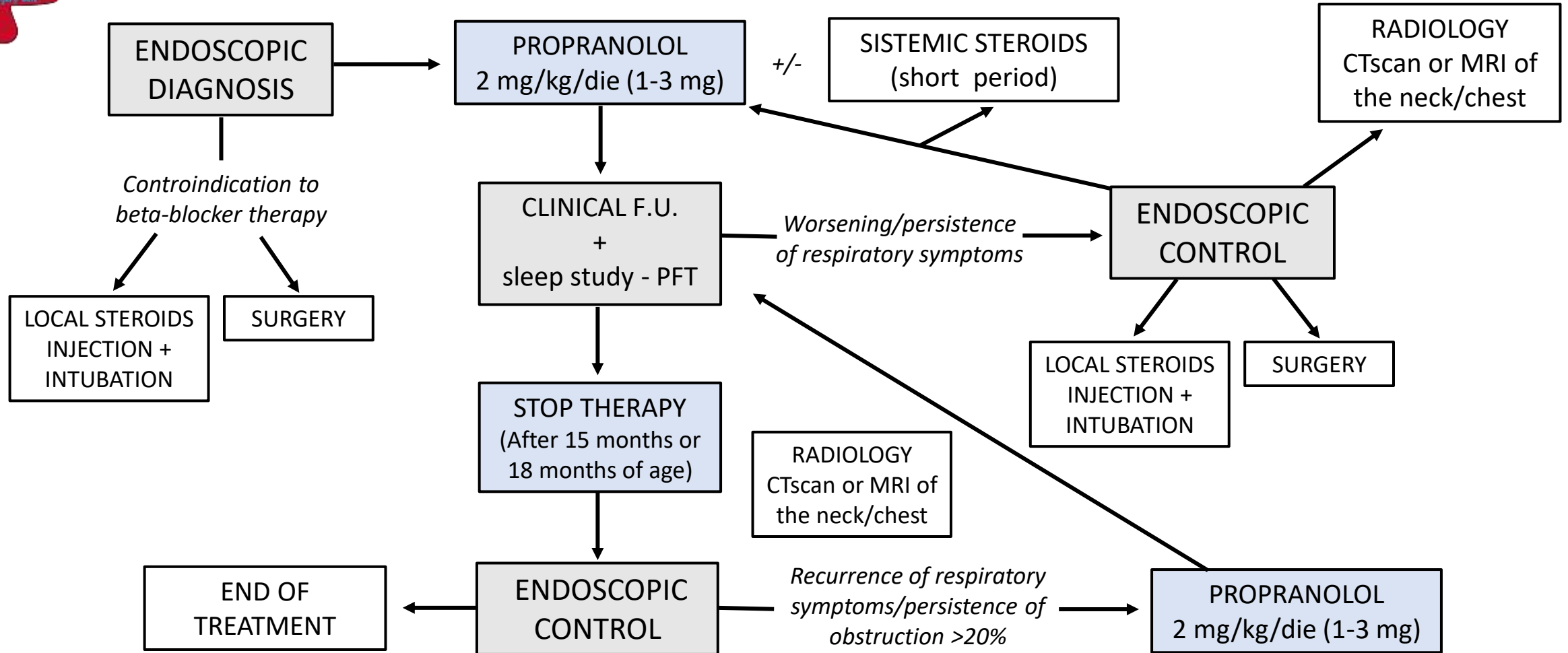
## ENDOSCOPY

SG obstruction **before therapy** – 10-80% (average 50,29%)

SG obstruction **after therapy** – 0-30% (average 7%)

> **eccentric localization** pattern (64.7% of cases)

# Our Protocol





# Our Protocol

## 1. ENDOSCOPIC DIAGNOSIS

(MRI or CT scan only for cases resistant to therapy, differential diagnosis or PHACES syndrome)

## 2. START OF THERAPY WITH PROPRANOLOL as soon as possible

(cardiological clearance for beta-blocker therapy)

2 mg/kg/die (1-3 mg/kg/ie)

## 3. SISTEMIC STEROIDS as adjuvant therapy based on clinical symptoms

## 4. CLINICAL FOLLOW-UP (consultation, sleep-study, PFT)

## 5. END OF THERAPY after 15 months or at 18 months of age

## 6. ENDOSCOPIC FOLLOW-UP only after 1 month from the suspension of therapy (except for worsening, persistence or recurrence of respiratory symptoms)



# Conclusions 1

- ✓ Laryngeal IHs could be **misdiagnosed** (the presentation is similar to other airway disease)
- ✓ The **high efficacy of  $\beta$ -blockers** has radically changed the prognosis
- ✓ **Surgery** is now indicated for difficult, refractory cases or for absolute contraindications to  $\beta$ -blockers
- ✓ **Delay of diagnosis and treatment** can lead to a failure of propranolol therapy
- ✓ **Several protocols** for the management of airway IHs have been proposed, but none has been standardized so far

*Elluru, R. G. et al. Multicenter Evaluation of the Effectiveness of Systemic Propranolol in the Treatment of Airway Hemangiomas. Otolaryngol. - Head Neck Surg. 153,452–460 (2015)*

*Perkins, J. A., Chen, E. Y., Hoffer, F. A. & Manning, S. C. Proposal for staging airway hemangiomas. Otolaryngol. - Head Neck Surg. 141, (2009)*

*Balakrishnan, K. & Perkins, J. A. Management of airway hemangiomas. Expert Review of Respiratory Medicine vol. 4 455–462 (2010)*



## Conclusions 2

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✓ Based on our experience:

- An early diagnosis is crucial
- Start the therapy with Propranolol as soon as possible
- Stop the therapy after 15 months or at 18 months of age
- Reserving endoscopy only for the diagnosis and the follow-up after the end of therapy
- Clinical follow-up is mandatory, also after the end of the therapy to promptly manage possible relapses

**Grazie per  
l'attenzione**



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