

## VIII SISAV International Congress

Vascular anomalies:  
update on diagnostic  
and therapeutic approach

Roma

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# A rare case of infantile hemangioma associated with pelvis syndrome

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# Background

- ✓ Infantile hemangiomas are the most common *benign vascular tumors* of infancy.
- ✓ Classification (ISSVA): focal, multifocal, segmental and indeterminate according to their distribution, and by its depth into superficial, deep or mixed.
- ✓ Infantile hemangiomas usually present before the first 4 weeks of life, and complete most of their growth by 5 months of age, it may extend up to the first year of age. Involution starts between 6 and 12 months and extends up to around 4 years of age in most patients however, residual skin changes are common.
- ✓ In addition to the cosmetic consequences, it is well established that segmental infantile hemangiomas can be associated with developmental structural anomalies.
- ✓ In some cases hemangiomas may be related to syndromes including PHACE, LUMBAR and **PELVIS syndrome** [1].



# PELVIS SYNDROME

- **P**erineal hemangioma
- **E**xternal genitalia malformations
- **L**ipomyelomeningocele
- **V**escicorenal abnormalities
- **I**mperforate anus
- **S**kin tag



Not all the components of the acronym are necessary to fit the malformation sequence

# CASE

Patient age: 4 yo, Gestational age: 41w

Echocardiography: FOP

No other imaging.

- Diagnosis at birth of MAR
- Colostomy at 3 days of life
- External genitalia characterized by:
  - normal left large lip
  - swelling of the right large lip with purple spots (malformation vascular?)
  - presence of vaginal ostium urethral ostium displaced to the right
  - recto-vulvar fistula.



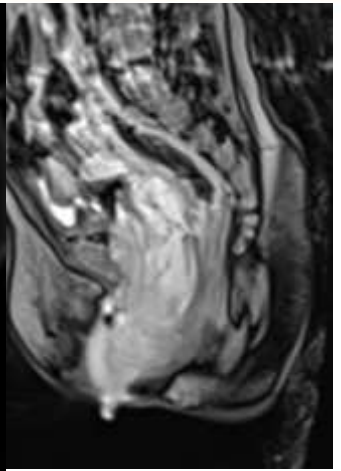
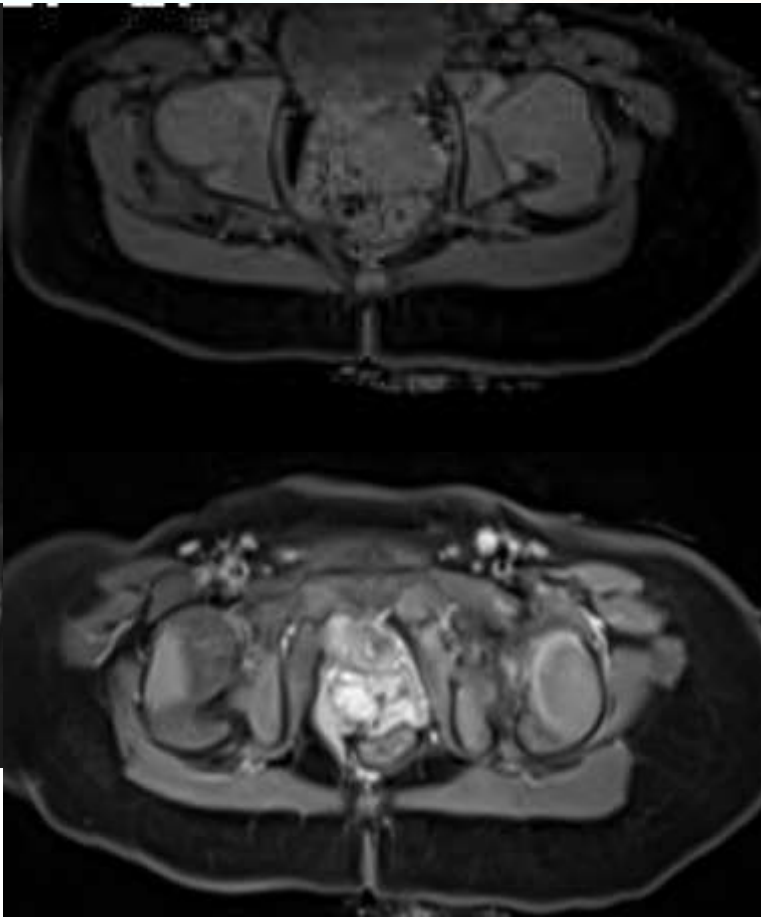
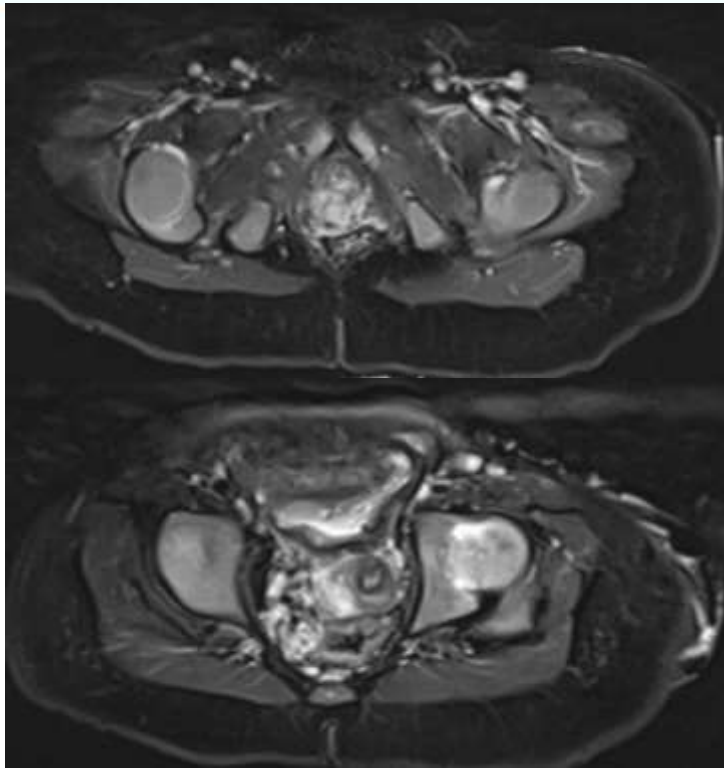
# PELVIS SYNDROME

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- **V**
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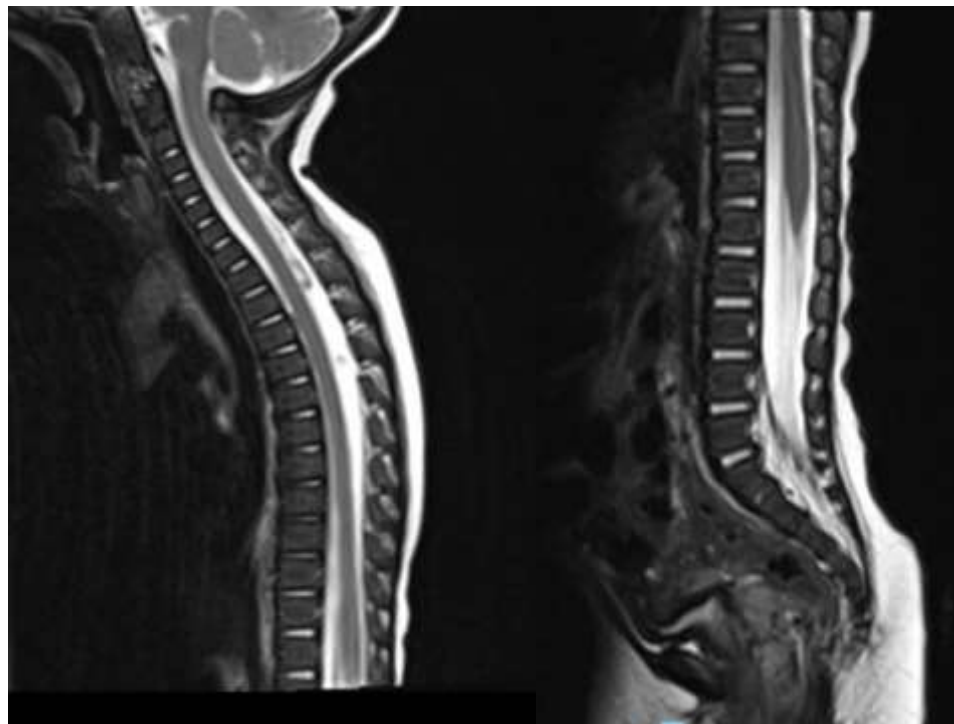
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*Inhomogeneous solid tissue that envelops the entire vaginal canal; caudally, the tissue reaches the perineal plane, with further tubular structure located anterior and closely to the tissue.*



No spinal  
dysraphism

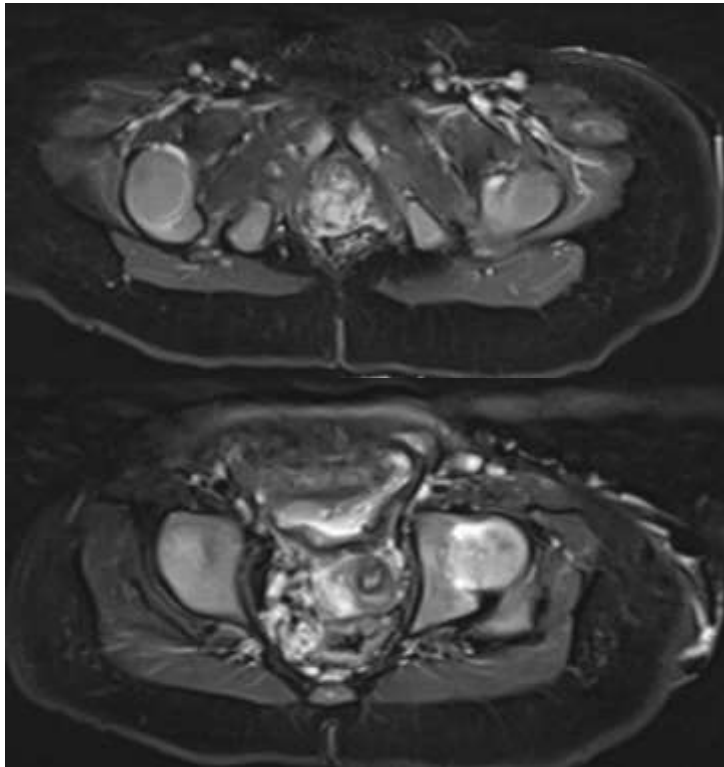
Anamnesis, clinical and radiological findings are referable to vascular anomaly as *segmental anogenital hemangioma*.

A therapy with **propranolol** was assessed

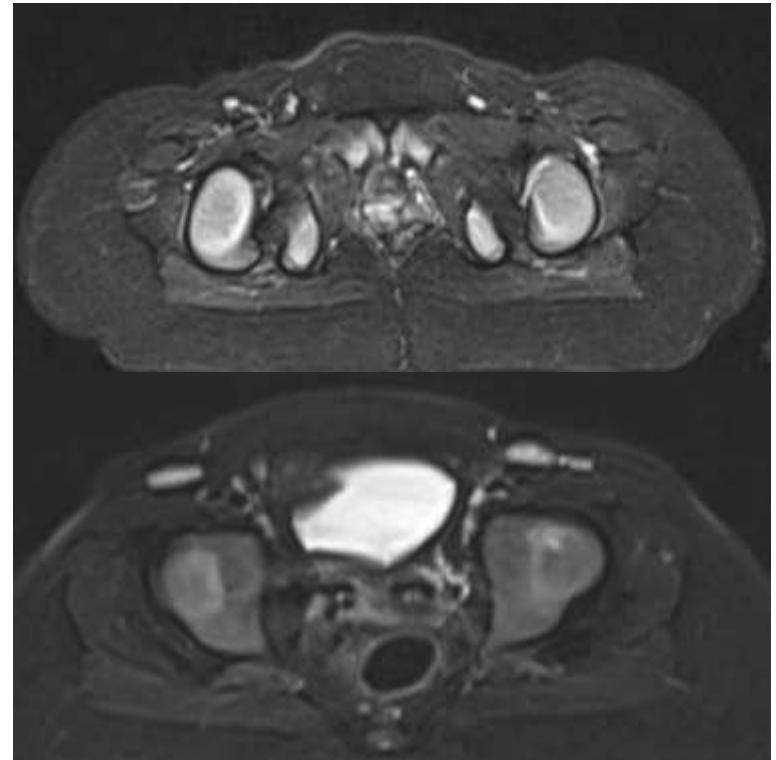
Two months later another abdominal MRI was performed and **solid tissue was reduced.**



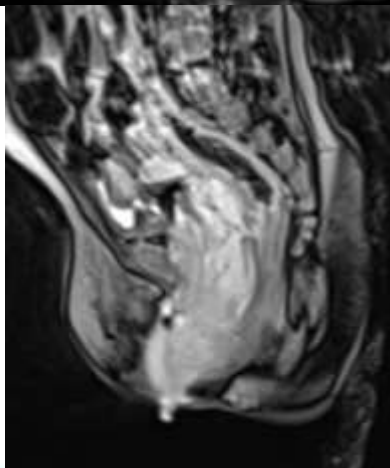
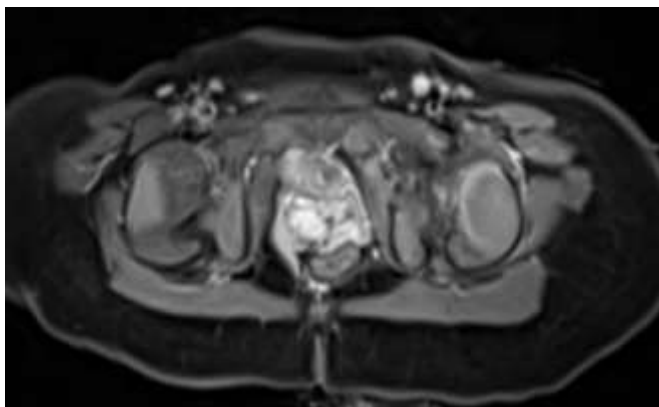
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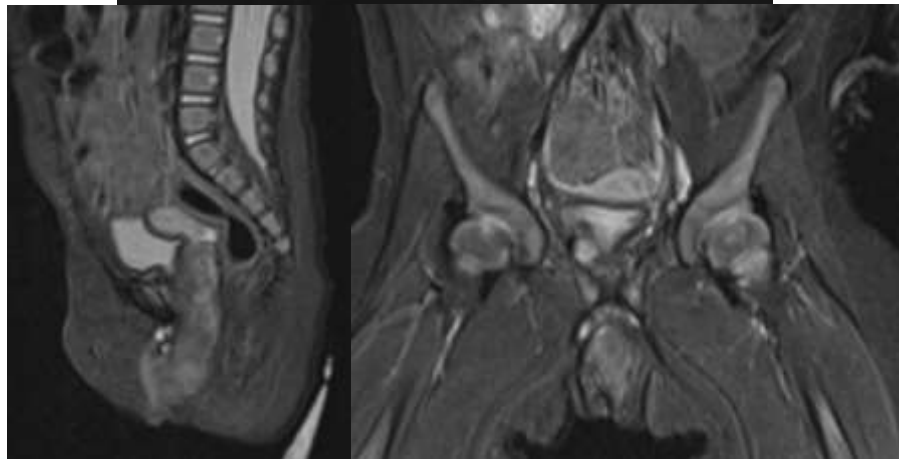
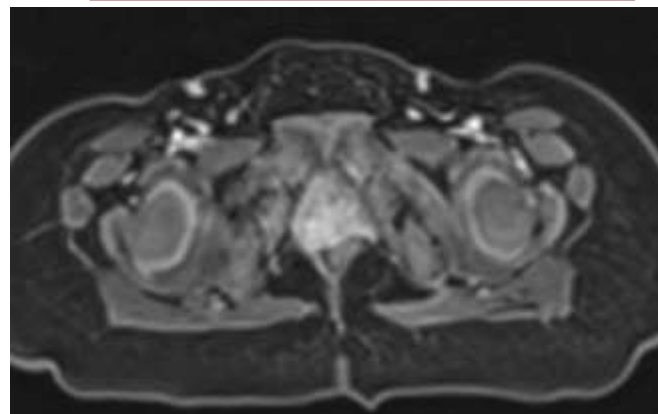
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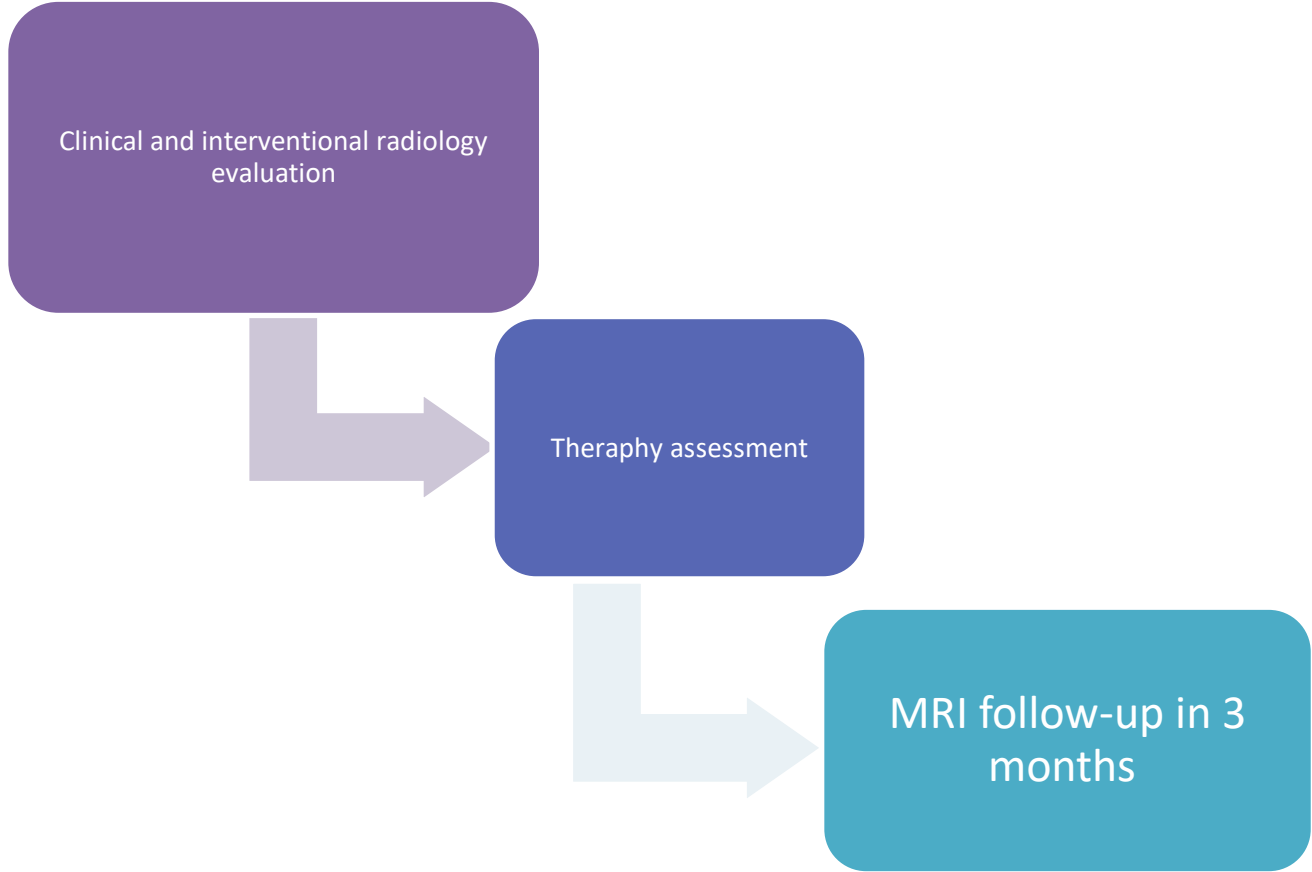


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# Take home messages

- ✓ In case of lesions with signal findings similar to hemangioma but with an atypical anatomic location, however think to infantile hemangioma
- ✓ MRI protocol: multiparametric/fat sat
- ✓ In these case (often MRI sedation) think to exclude a syndrome and associated malformations (ex spinal malformation)



# Thanks for your attention

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