

PERSPECTIVES 2017

December Friday 15 - BORDEAUX

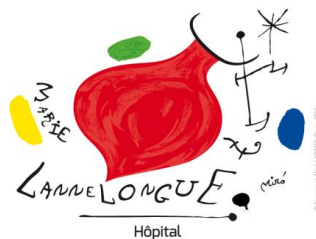
Organization: E. Ducasse, M. Sibé



www.congresperspectives.com

Ischémie critique à 20 ans: Méfiance...

**Dominique FABRE, Antoine GAUDIN, Thomas Le HOUEROU, Come BOSSE,
Alessandro COSTANZO, Stephan HAULON.**



HOPITAL MARIE LANNELONGUE
Groupe Hospitalier Paris Saint-Joseph
UNIVERSITE PARIS-SUD SACLAY



Disclosure of Interest



Speaker name: Dominique Fabre

- I do not have any potential conflict of interest with this presentation

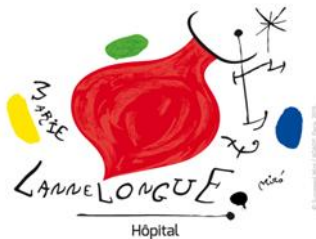
CLI during Sars Cov-2

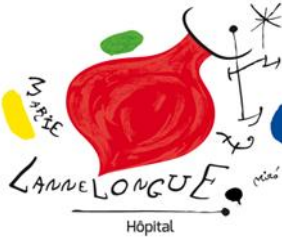
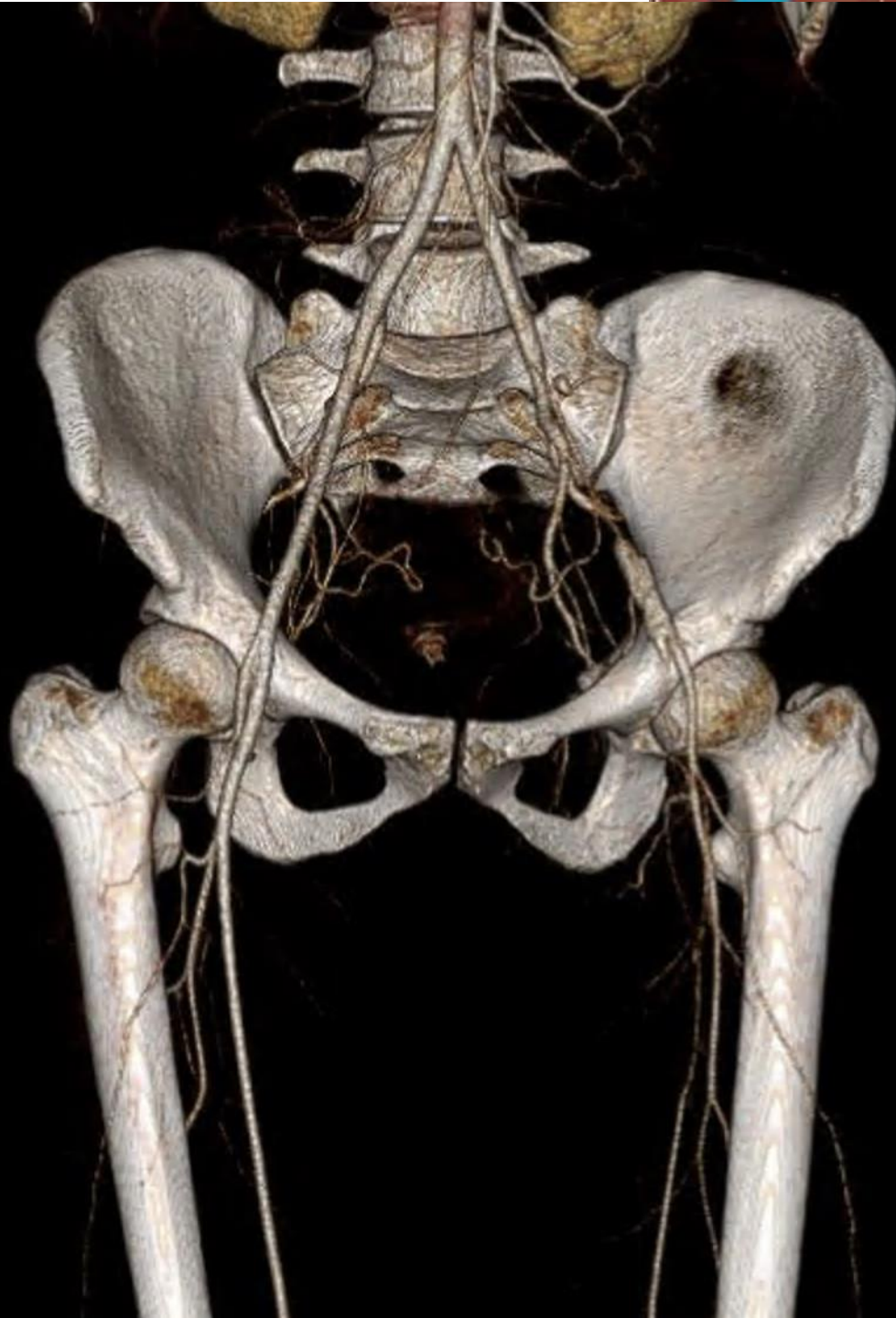
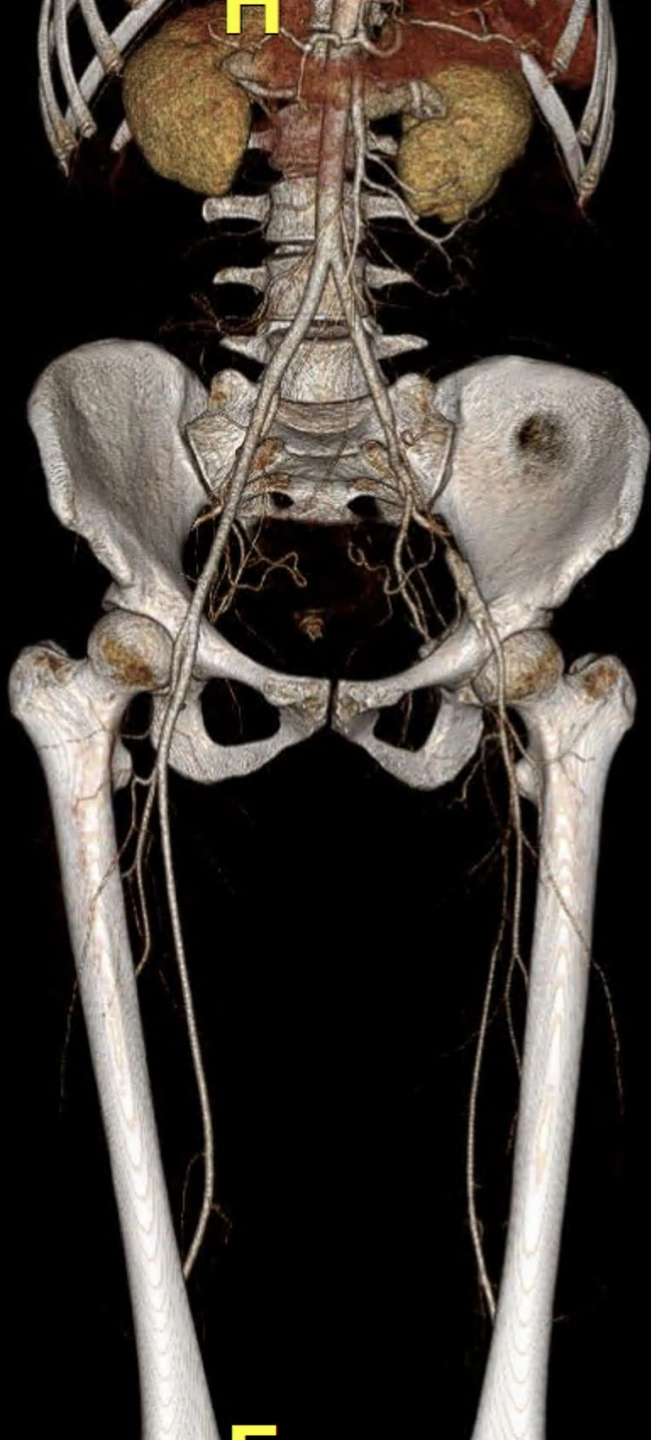
20 Years old W, no smoking, good radial pulse
CLI since few weeks 15 j
Rest pain / Sensitive ischemia leg and feet / partial motor

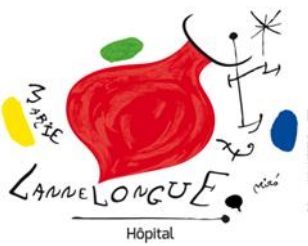
CT SCAN:

Spontaneous dissection left external iliac artery.
Dissection left common femoral artery gauche
occlusion proximal part SFA
Small patent profunda

HEPARIN / ASPIRIN

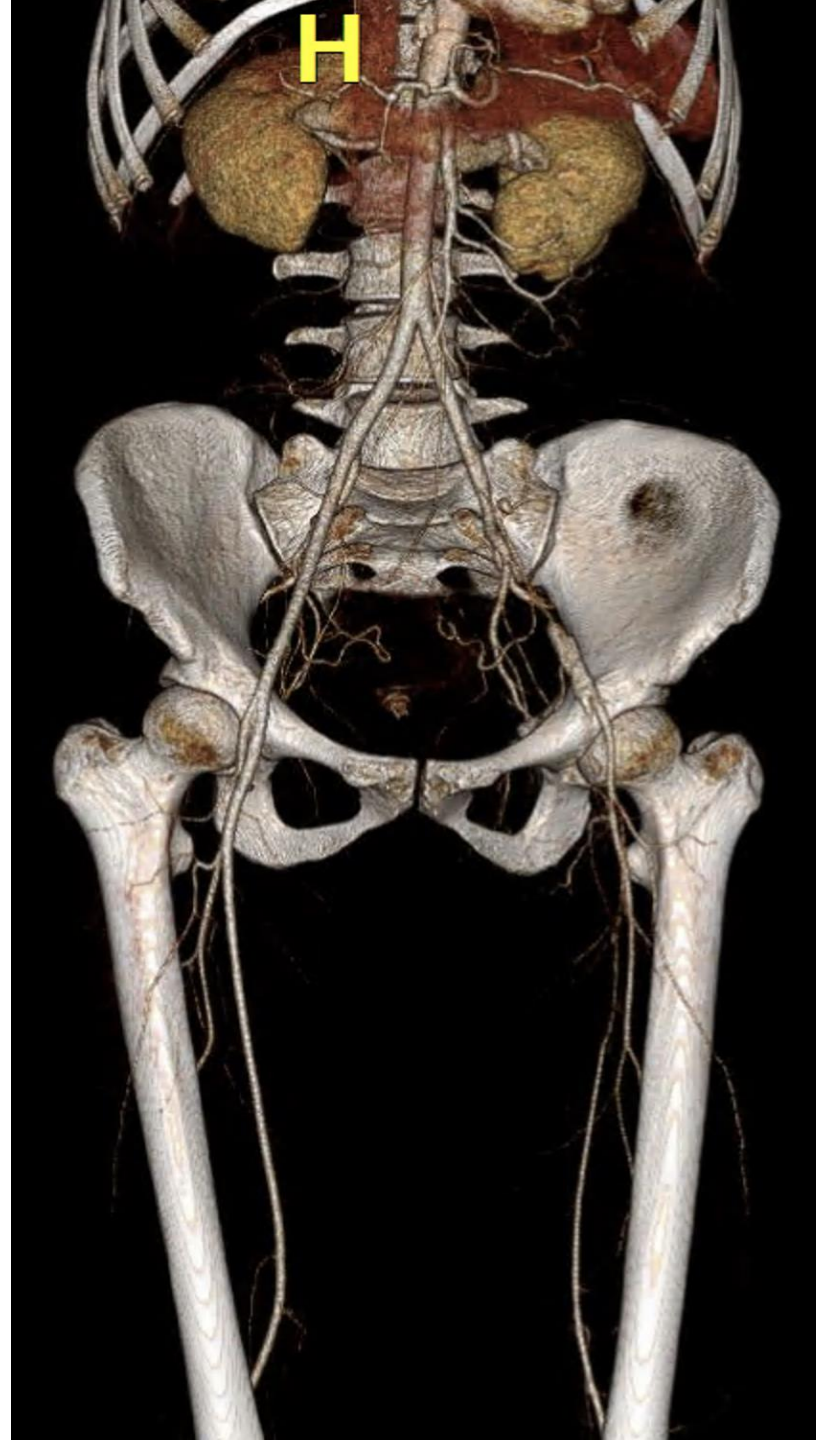
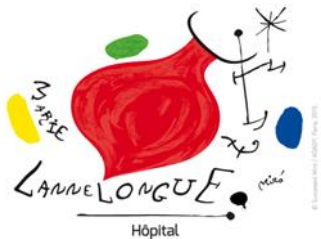




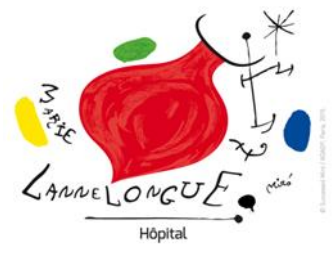


Biology

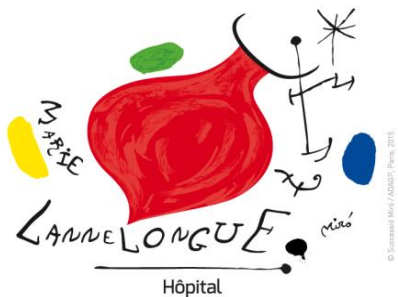
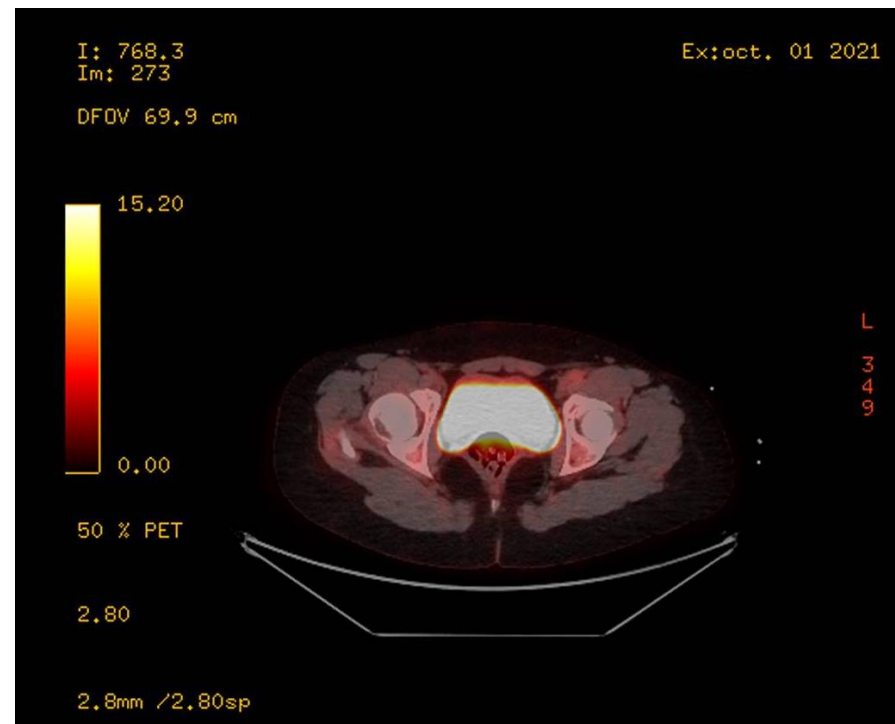
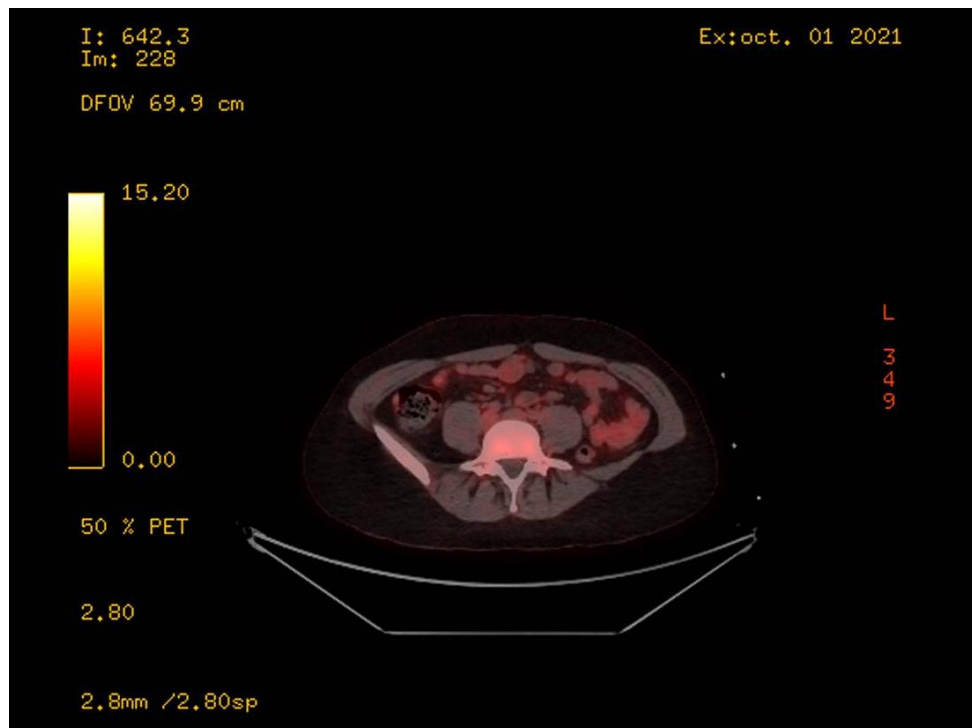
- Creatinine 42 $\mu\text{mol.l}$
- Fibrinogène 3.4 g/l
- Leucocytes 11300 / mm^3
- Hb 11 g/l
- Plaquettes 360000



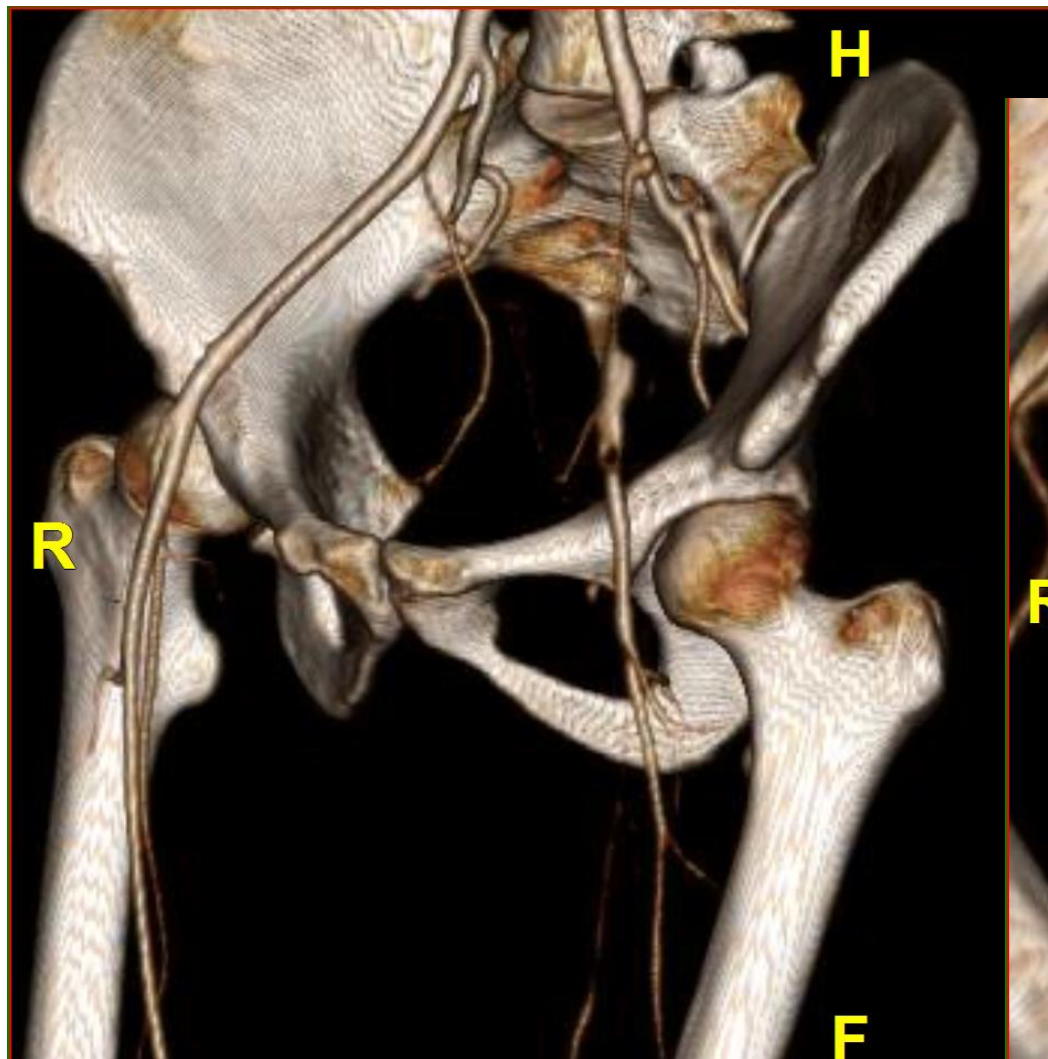
PET CT scan



PET CT



CT SCAN



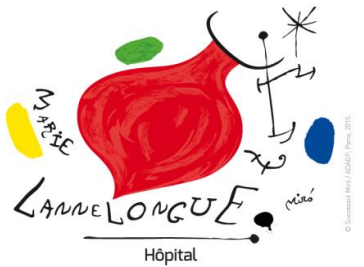
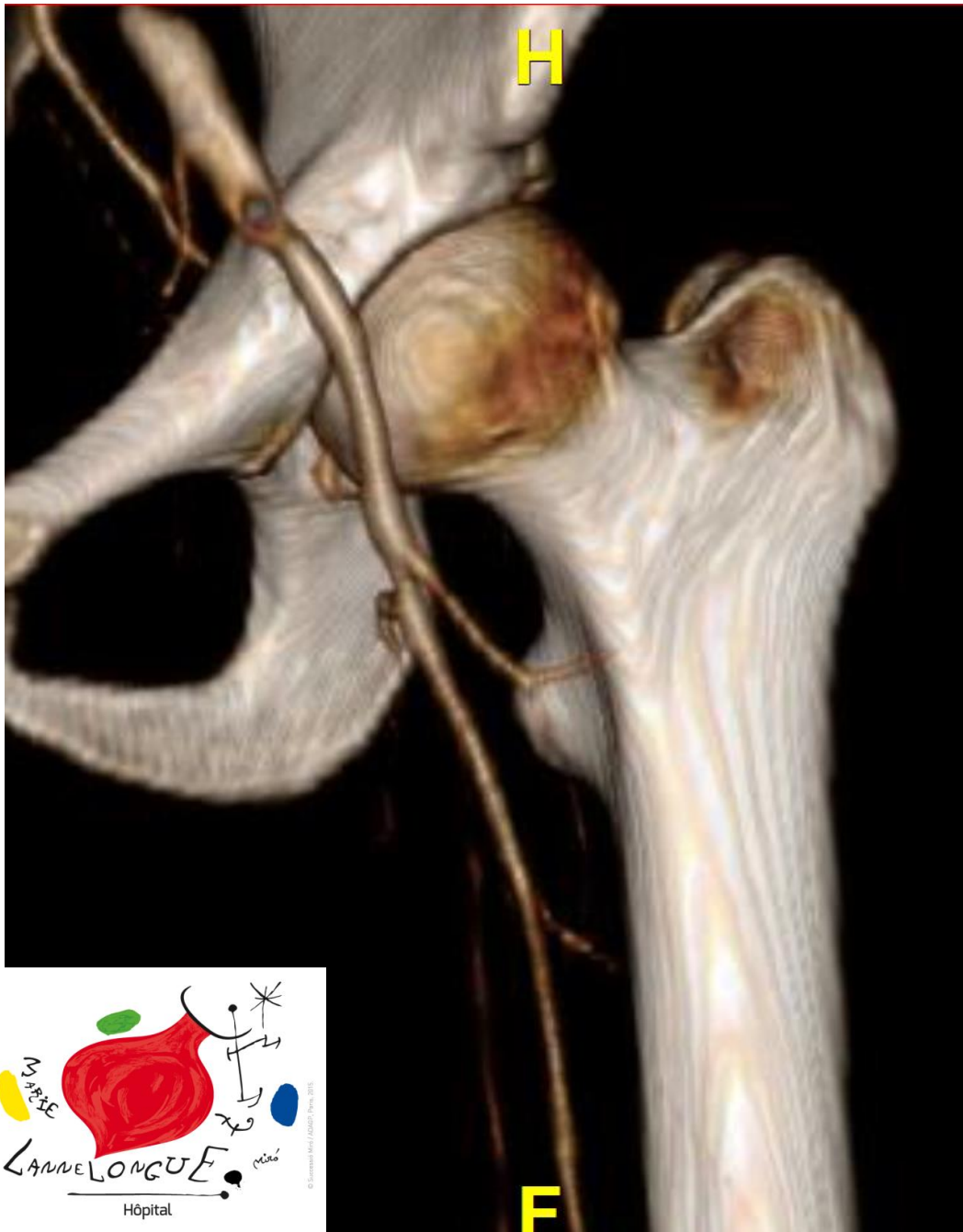


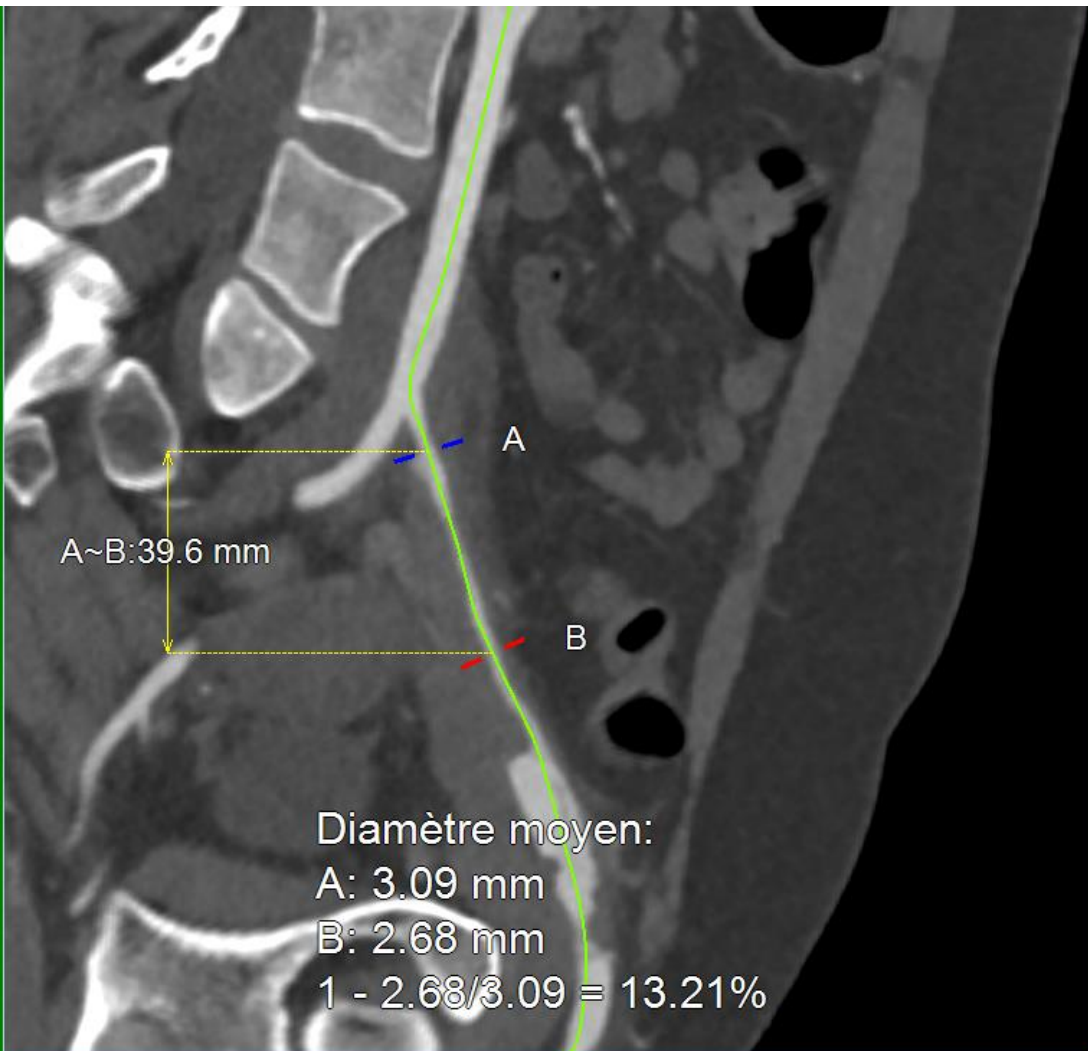
5m



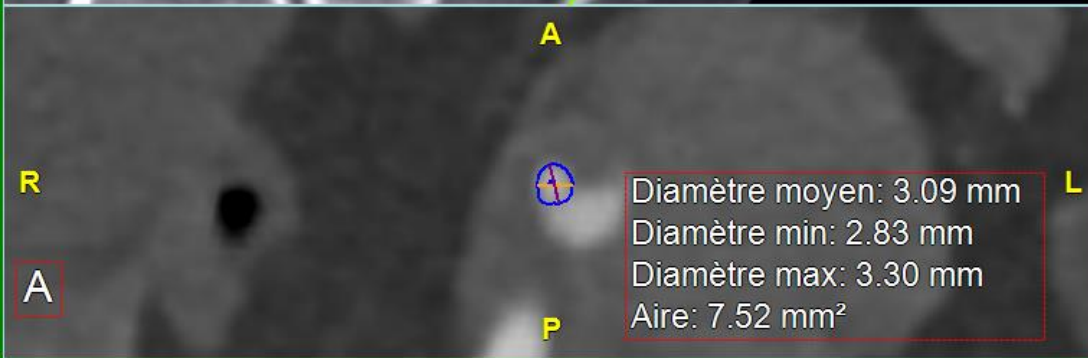
Comprendre le monde,
construire l'avenir







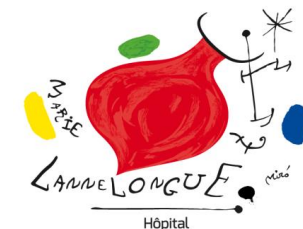
Diamètre moyen:
 A: 3.09 mm
 B: 2.68 mm
 $1 - 2.68/3.09 = 13.21\%$



Diamètre moyen: 3.09 mm
 Diamètre min: 2.83 mm
 Diamètre max: 3.30 mm
 Aire: 7.52 mm²



Diamètre moyen: 2.68 mm
 Diamètre min: 2.5 mm
 Diamètre max: 2.77 mm
 Aire: 5.66 mm²



Clinical evolution

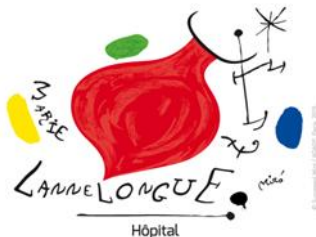
No improvement under Heparin

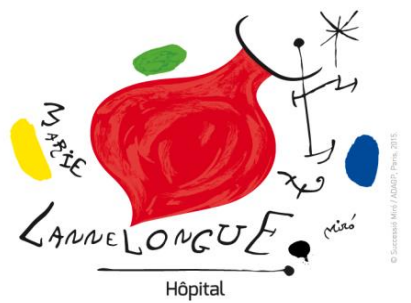
No walking possibility

CLI

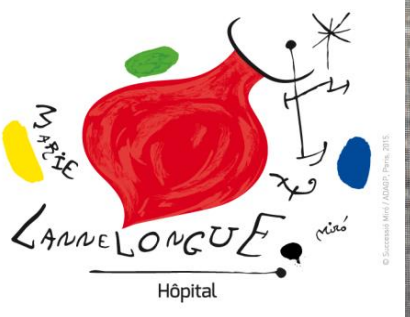
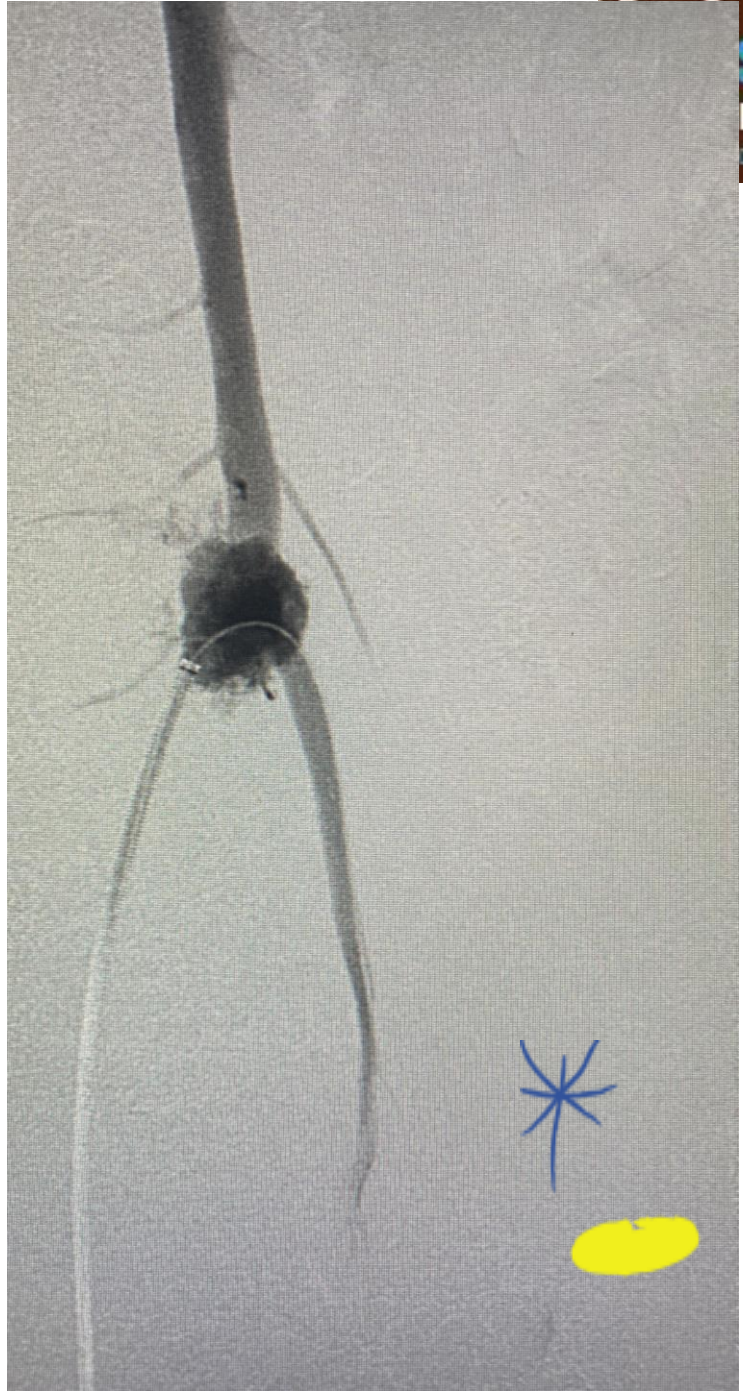
Before completing the treatment

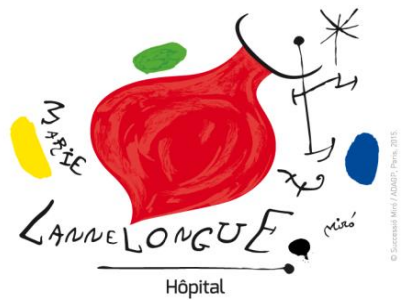
Decision to start with an endo approach











© Eurocard Miro / ADAGP Paris, 2015.



B O R D E A U X
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Organization: E. Ducasse, M. Sibé



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SUD**

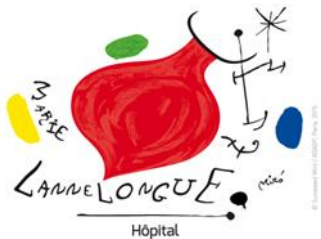
Comprendre le monde,
construire l'avenir

université
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Aortic rupture

- Hemorrhagic choc
- Complete rupture of the distal aorta
- Disjunction of both iliac
- Laparotomy
- Aortic + iliac clamping
- Anterior rupture IVC

- General anesthesia
- Arterial catheter
- Central venous access




Open repair

- Rupture external iliac, common femoral, profunda, SFA
- **Aorto popliteal bypass**
- **Reimplantation right iliac**
- Ligation of all arteries
- Distal anastomosis popliteal artery



- Nad 8 Mg /h
- Lactates 9
- Remplissage 8 L
- 13 Culots globulaires
- 12 PFC
- 4.2 l cell saver
- 9g fibrinogene
- 10 CPU / plaquettes

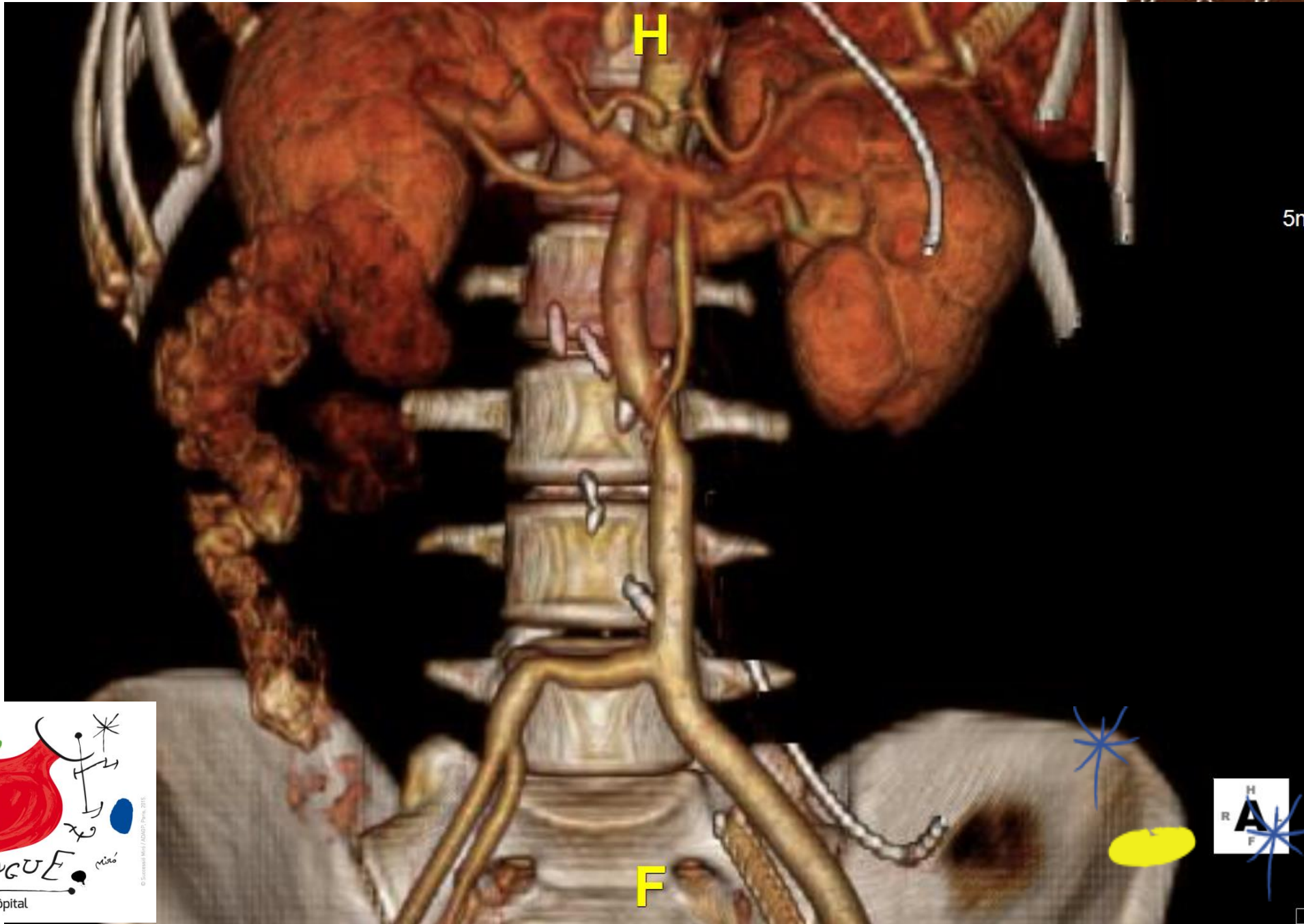


Angioplasty timing / 8 h

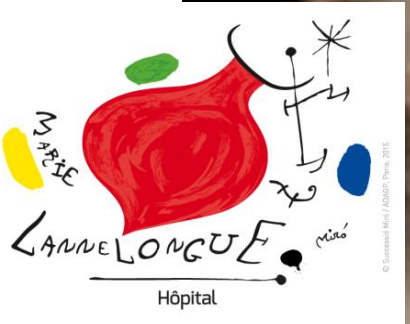


ENTREE EN SALLE	11:58
DEBUT DE LA	12:32
FIN DE LA PROCEDURE	18:30
SORTIE DE SALLE	19:46



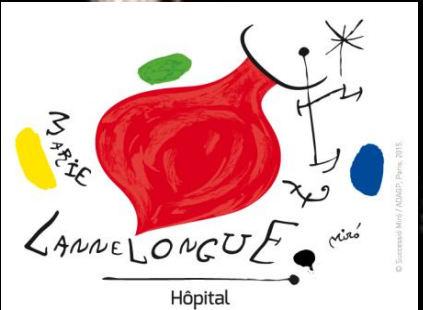
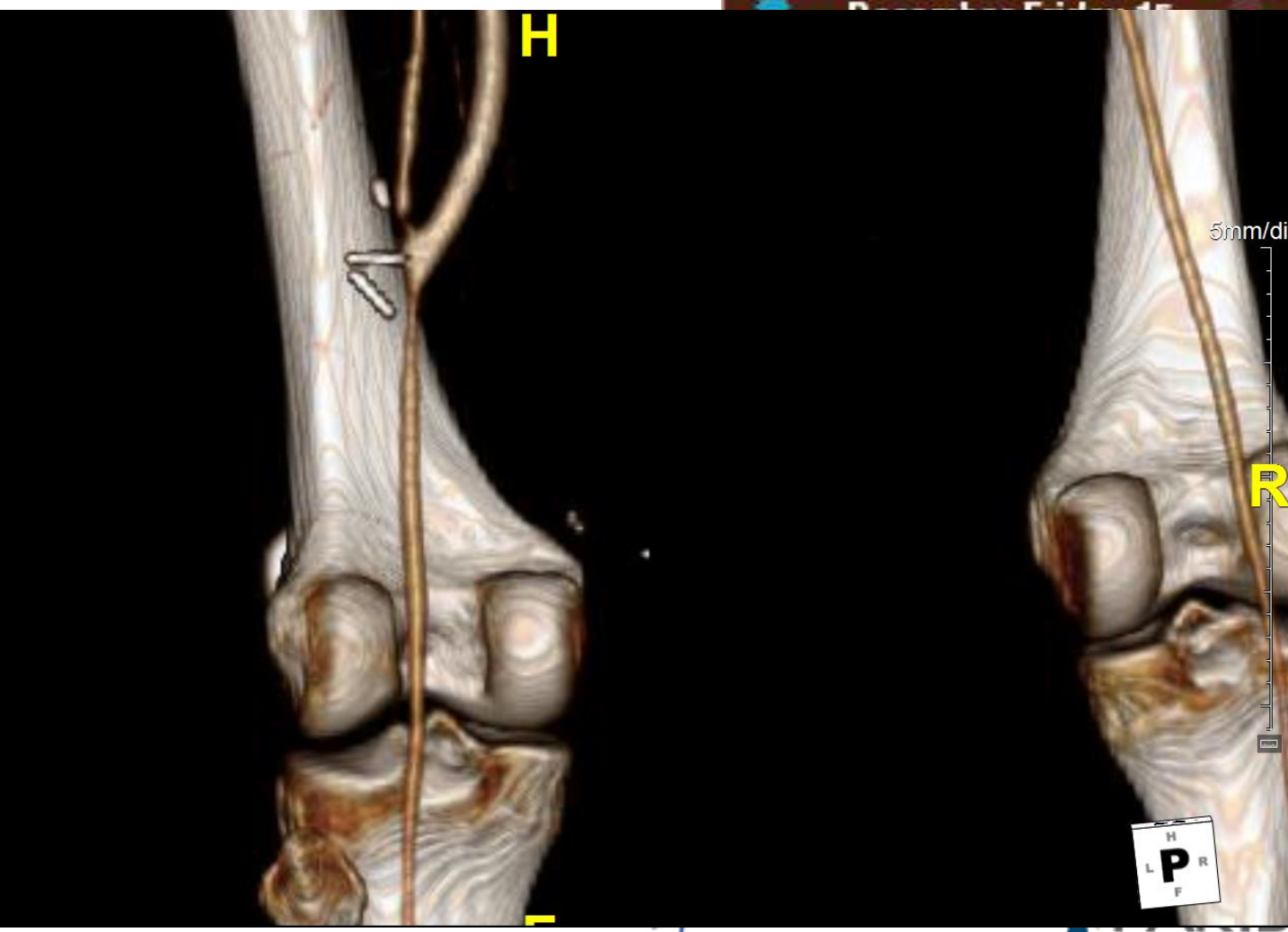
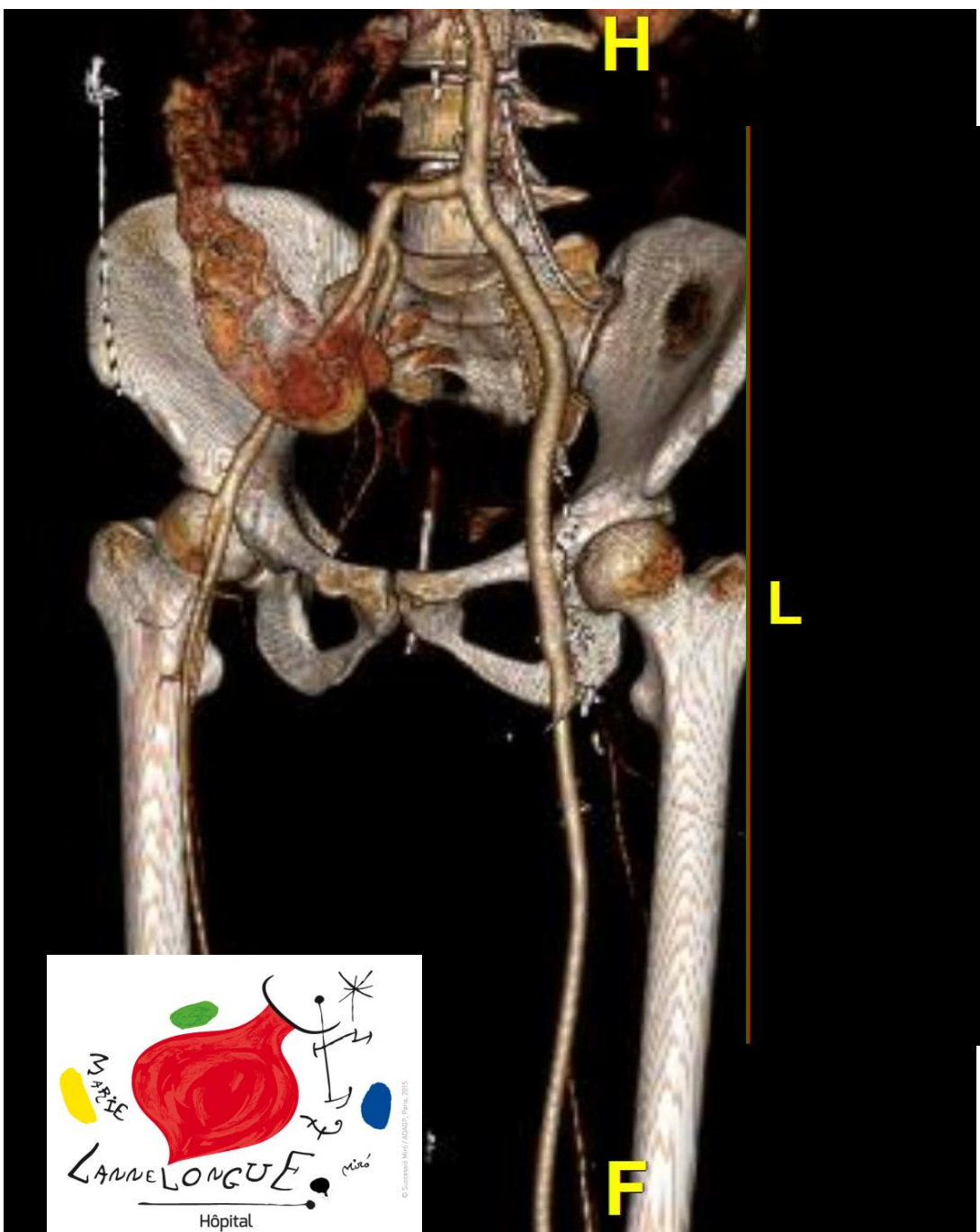


5n



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construire l'avenir





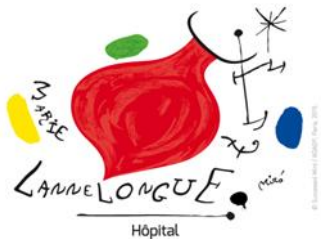
Out of hospital in 2
weeks



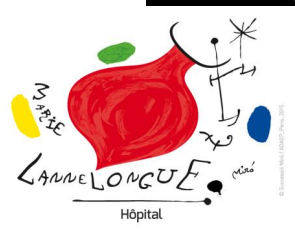
Follow-up 1 month



- Back pain
 - Fever
 - Inflammatory markers
 - Suspicion of infection
-
- CT SCAN



A



A
R E



Comprendre le monde,
construire l'avenir





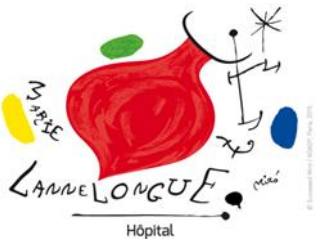
SA
Saclay

Hôpital

Double JJ placement

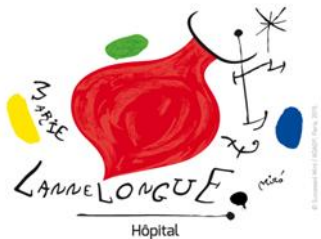


- Bacteriuria
- Complete recovery after double J placement



Follow up at 2 months

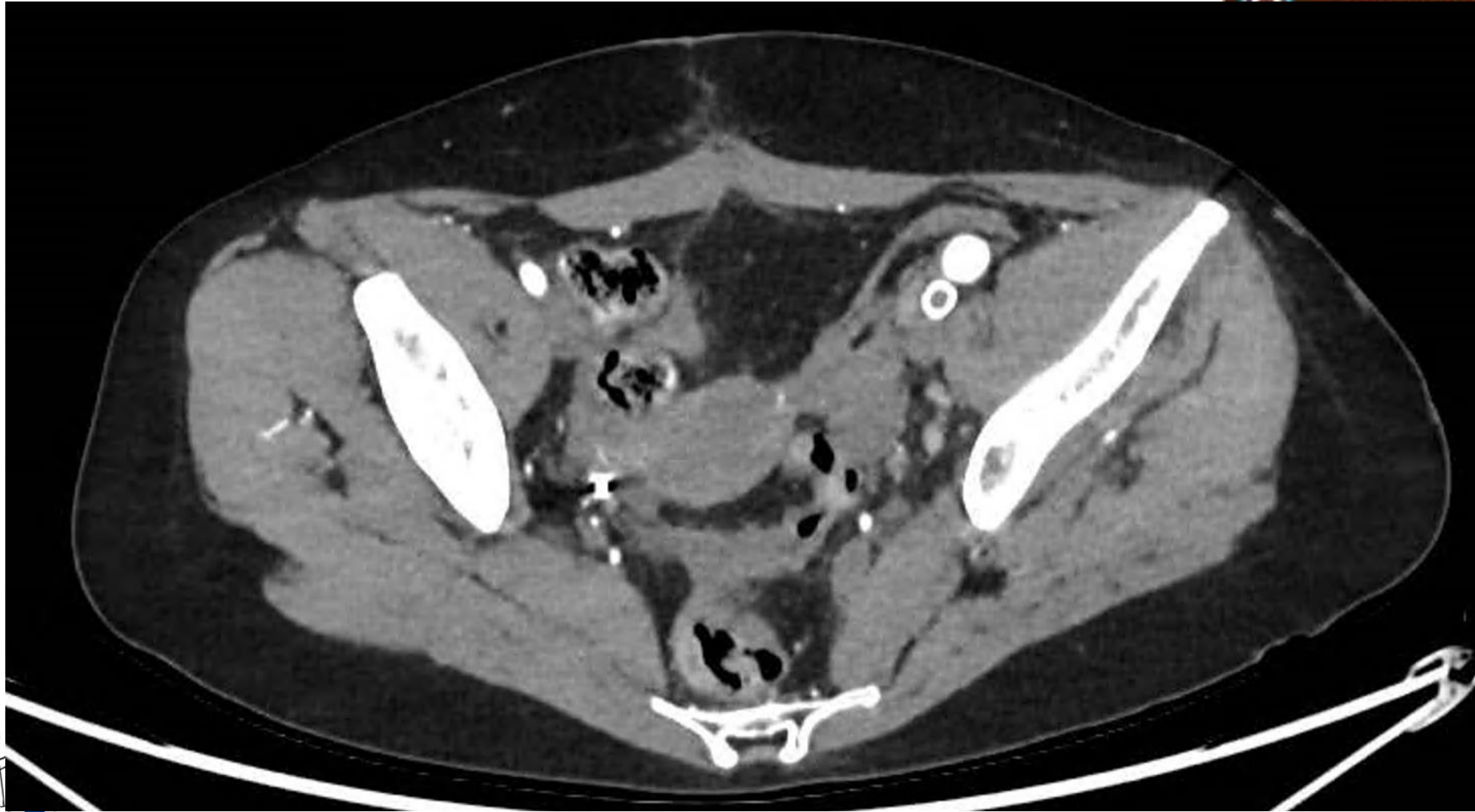
Fever
Inflammatory markers
Suspicion of infection

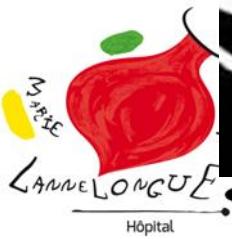
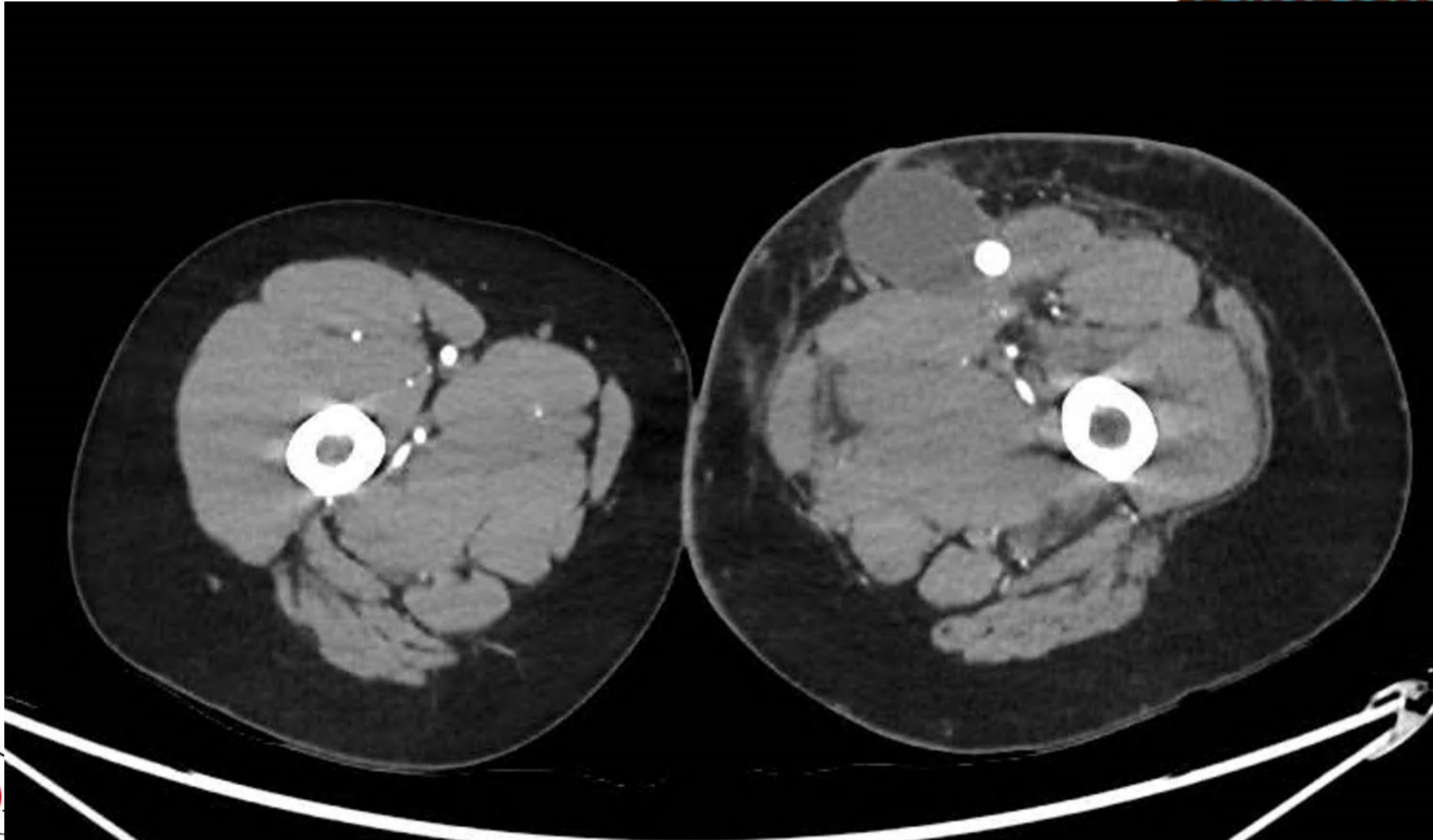


Follow up at 2 months







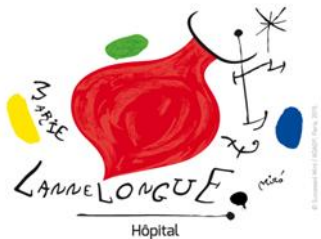


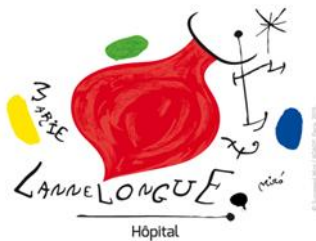
Comprendre le monde,
construire l'avenir



Operation

- Subcutaneous infection
- No proximal extension
- No abdominal infection





DIAGNOSTIC CRITERIA FOR EHLERS-DANLOS SYNDROME, VASCULAR TYPE

MAJOR DIAGNOSTIC CRITERIA

- Thin, translucent skin
- Arterial/intestinal/uterine fragility or rupture
- Extensive bruising
- Characteristic facial appearance (thin delicate nose, thin lips, hollow cheeks)

MINOR DIAGNOSTIC CRITERIA

- Acrogeria (taut, thin skin)
- Hypermobility of small joints
- Tendon and muscle rupture
- Talipes equinovarus (clubfoot)
- Early-onset varicose veins
- Arteriovenous, carotid–cavernous sinus fistula
- Pneumothorax/pneumohemothorax
- Gingival recession
- Positive family history, sudden death in one or more close relatives

Data from Beighton P, et al. Ehlers-Danlos syndromes: revised nosology, Villefranche, 1997. *Am J Med Genet* 77:31-37, 1997.

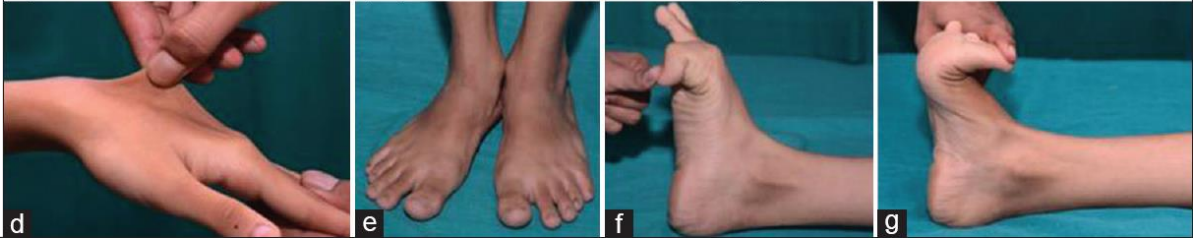
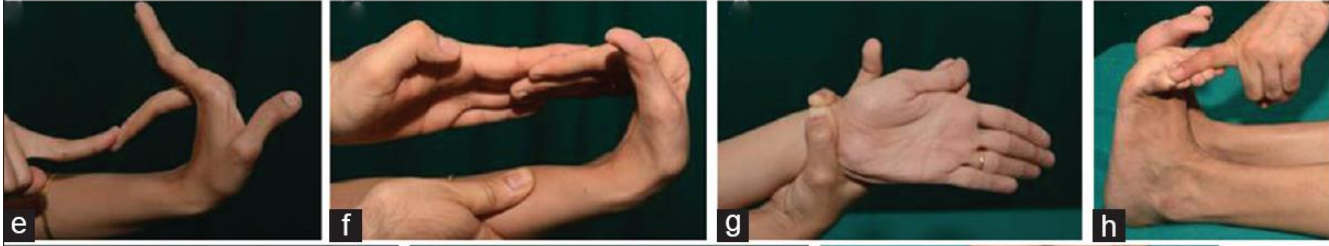


CRITERES MAJEURS

Peau fragile fine et translucide
Rupture: artérielle, intestinale,
utérine

Ecchymoses extensives

Morphotype facial: nez fin, lèvres
fines, Joues creuses



Vascular = Type IV

Table 143-2		Subtypes of Ehlers-Danlos Syndrome				
Nomenclature (New Terms)	Type	Skin (0-4+) (Elastic-Fragile)	Joint Laxity (0-3+)	Features	Inheritance	
Classic	I, II	+++ / +++	+++	Vascular complications rarely	AD	
Hypermobile	III	+ / +	+++	Arthritis	AR	
Vascular	IV	- / ++++	+	Rupture of arteries, uterus, intestine; thin skin	AD	
Kyphoscoliotic	VIA, VIB	+++ / ++	+++	Hypotonia, osteoporosis, kyphoscoliosis; rupture of arteries, globe of eye	AR	
Arthrochalasic	VIIA, VIIB	++ / +	+++	Hip subluxation, osteoporosis	AD	
Dermatosparactic	VIIIC	- / ++++	+	Skin doughy and lax	AR	
Other	V	++ / ++		Skin lax	X-linked	
	VIII	+ / ++	++	Periodontal disease	AD	
	IX	+ / -	+	Lax skin, osteoporosis, bladder diverticula, retardation	X-linked	
	X	+ / +	++	Petechiae	?	

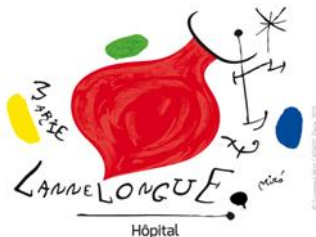
AD, Autosomal dominant; AR, autosomal recessive.

Prevalence EHLERS DANLOS type IV

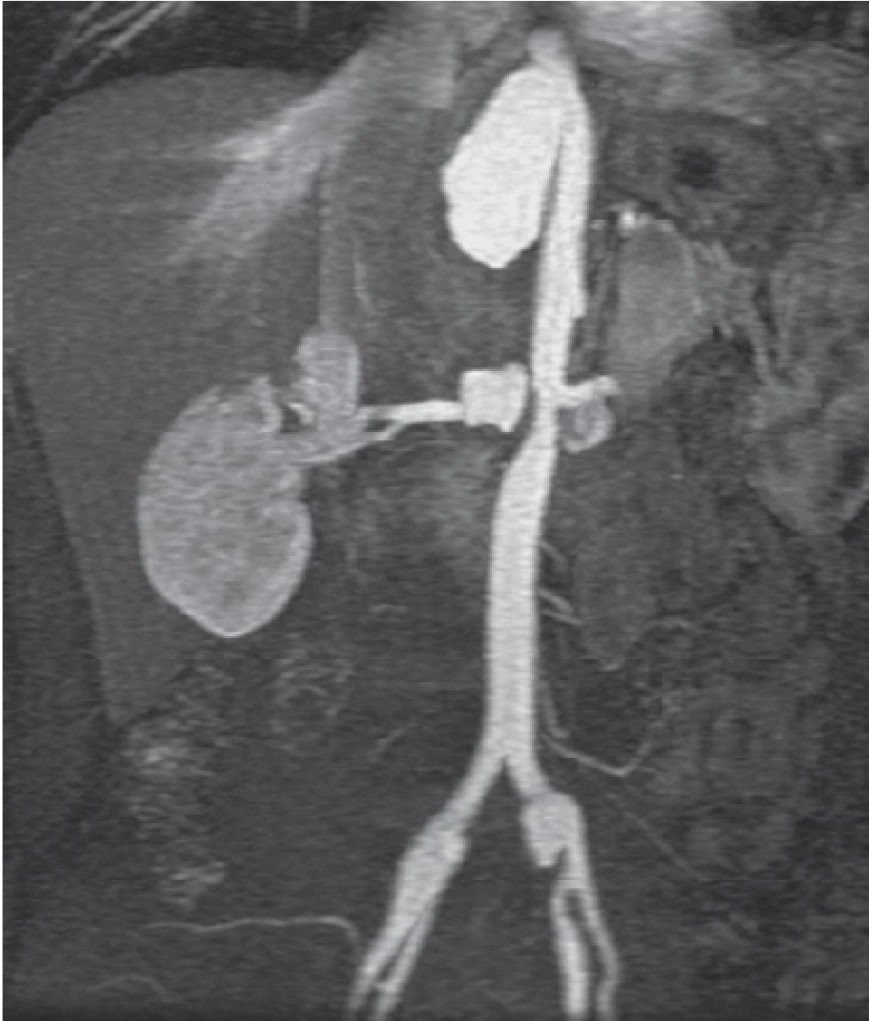


- Environ 500 cas repertoriés en France
- 1 / 150 000 patients

- Premier cas d'hyperlaxité décrit par Hippocrate 400 avant JC
- 1900: Société danoise Dermatologie , Edvard EHLERS, cas hyperlaxité et atteinte cutanée
- 1908: Henri-Alexandre DANLOS, Société Française Dermatologie , peau minceur anormale
- 1933: Classification ED / syndrome cutanéoligamentaire
- 2017: new classification / 13 sous types



Ehlers Danlos Syndroma

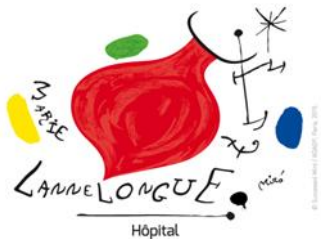


50% of new mutations that
appears sporadically
Mutation in COL3A1 Gene
Code for procollagene III

Post op treatment

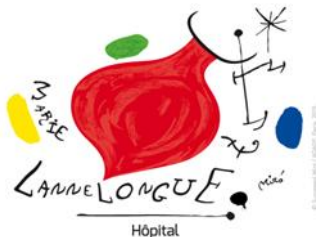


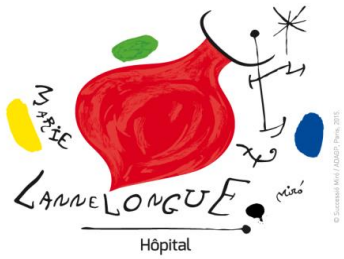
- Aspirin
- Bblockers
- Psychologist
- Protection / life



Genetic testing at 6 months

- Double cutaneous biopsy
- RNA transcript analysis
- Mutation in COL3A1 Gene
- Code for procollagene III:
 - Variation class 3 ACMG 2015 / COL3A1 gene / alteration epissage intron 36





Conclusions

Le syndrome d'Ehlers Danlos vasculaire type IV est rare et nécessite en cas de suspicion de prévoir un geste chirurgical de recours.

La rupture aortique peut survenir lors d'un geste endovasculaire.

L'espérance de vie d'un patient avec un syndrome d'Ehlers-Danlos type IV est de 48 ans, principalement du au rupture artérielle.

Le diagnostic génétique et la prise en charge se fait dans des centres de référence avec un délai diagnostique d'environ 6 mois.





C'est la profonde ignorance qui inspire le ton dogmatique. Celui qui ne sait rien croit enseigner aux autres ce qu'il vient d'apprendre lui-même; celui qui sait beaucoup pense à peine que ce qu'il dit puisse être ignoré, et parle plus indifféremment.

(Jean de La Bruyère)