

# Les fistules porto-systémiques congénitales

Emmanuel Gonzales, Stéphanie Franchi-Abella, Florent Guérin

# History

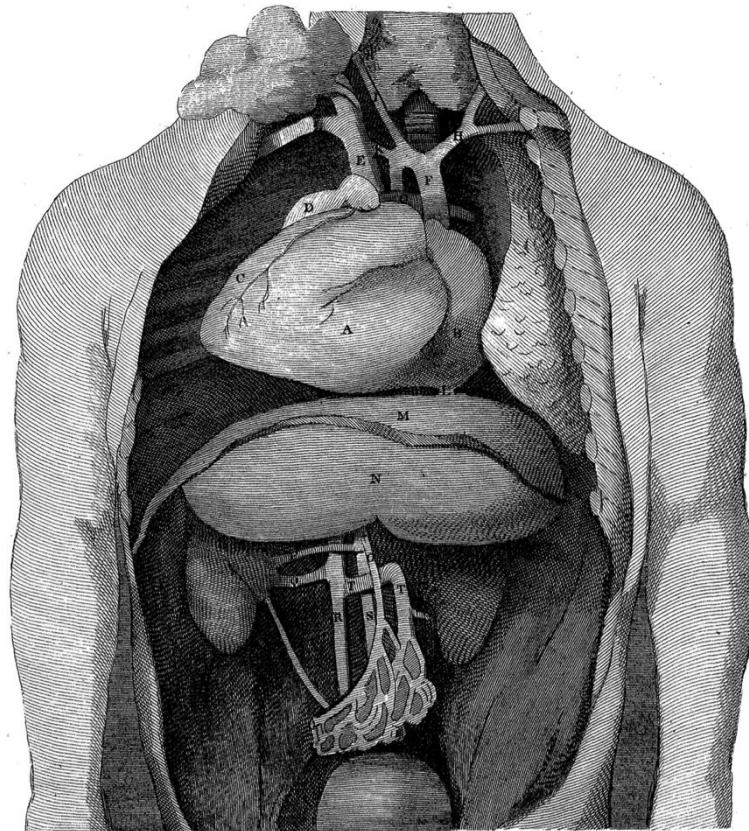
## First description by Dr John Abernethy 1793 , british surgeon

IX. *Account of two Instances of uncommon Formation, in the Viscera of the Human Body.* By Mr. John Abernethy, Assistant Surgeon to St. Bartholomew's Hospital. Communicated by Sir Joseph Banks, Bart. P. R. S.

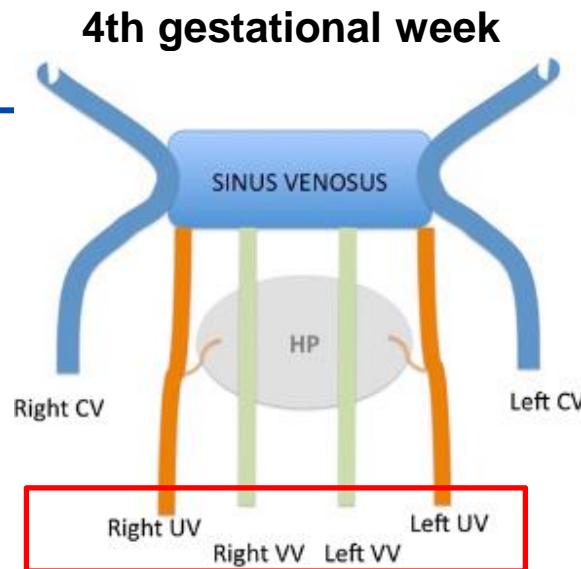
Read February 14, 1793.

I TAKE the liberty of presenting to the Royal Society, the relation of two cases of uncommon formation of the human body. When animal existence is supported by any other than the usual admirably contrived means, it cannot fail to excite the attention of the philosopher, since it shews to him the powers and resources of nature.

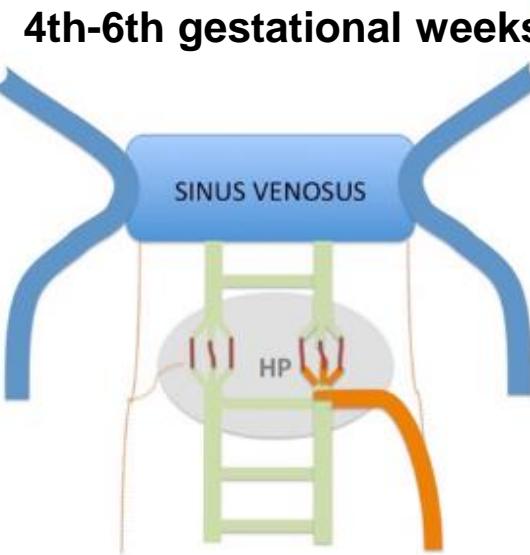
The peculiarities of the first case which I have the honour to offer to the Society, consist in an uncommon transposition of the heart, and distribution of the blood vessels; together with a very strange, and, I believe, singular formation of the liver. The body which contained these deviations from the usual structure was brought to me for dissection; with its history whilst alive, I am therefore unacquainted. The subject was a female infant, which measured two feet in length; the umbilicus was firmly cicatrized, and the umbilical vein closed; from these circumstances I conclude that it was about ten months old. The muscles of the child were large and firm, and covered by a considerable quantity of healthy fat; indeed the appearance of the body strongly implied that the child had, when living, possessed much vigour of constitution.



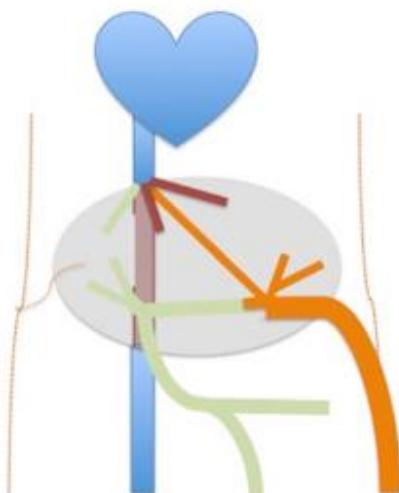
# Development of the hepatic vasculature



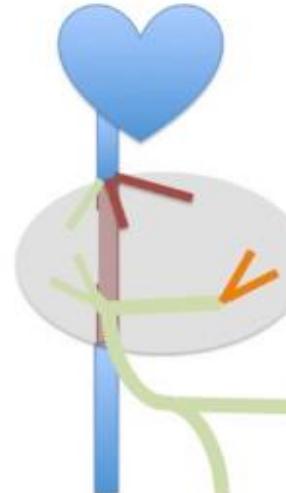
Extra-embryonic



Fetal circulation week 6



Newborn circulation



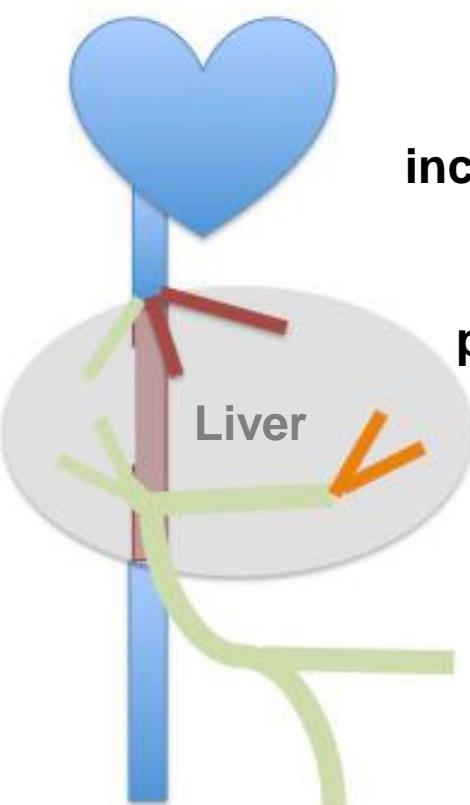
HP: hepatic  
primordium

# Definition of congenital porto-systemic shunt

**Congenital porto-systemic shunt:**

**incomplete vascular remodeling**

communication between  
**portal and systemic venous systems**



## Epidemiology:

- Newborn screen: 1:30'000
- Permanent: 1:50'000

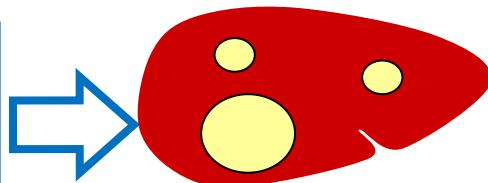
## Anatomical classification:

- Controversial
- Intrahepatic vs extrahepatic
- Portal vs no portal flow

## Newborn circulation

# Complications related to CPSS

portal  
deprivation  
of liver

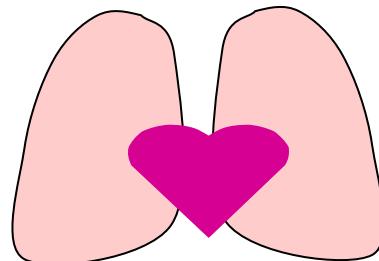


**Neonatal cholestasis,  
Cytolysis,  
Coagulation disorders**

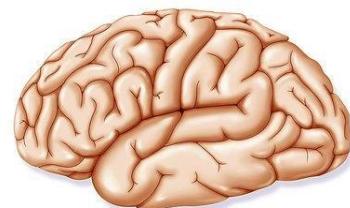
Focal Nodular Hyperplasia  
Adenomas

Regenerative Nodular Hyperplasia  
Hepatocellular Carcinoma

Splanchnic  
blood  
bypassing  
liver



Pulmonary hypertension  
A-V pulmonary shunts



Portosystemic encephalopathy  
**Hyperammonemia**

# Diagnosis

---

## Incidental finding

- Prenatal
- Post natal
- Adult

## Symptomatic

# Diagnosis: incidental finding

## ❖ Prenatal

- Umbilical Doppler
- Abnormal vessel size or number
- Anatomy difficult

## ❖ Post-natal

- Abdominal imaging for other indication
- At any age!

# Diagnosis: symptomatic

- **Clinically**
  - Liver nodule (s) or tumor(s)
  - Encephalopathy/neurocognitive
  - Cardiopulmonary complications
  - Other
- **Biochemically**
  - Normal LFTs
  - Cholestasis
  - Elevated plasma bile acids
  - ↑ galactose-1-phosphate (newborn screen +)
  - Hypoglycemia
  - ↑ plasma NH4<sup>+</sup>

# Symptomatic-Liver presentation

## ↗ Nodular liver

- Nodular regenerative hyperplasia
- Focal nodular hyperplasia
- Adenoma
- Hepatoblastoma
- Hepatocellular carcinoma
- Hemangioma

## ↗ Steatosis (hyperechoic liver on US)

# Symptomatic-heart and lung

---

- High output heart failure
- Hepatopulmonary syndrome
- Pulmonary hypertension

# Symptomatic-neurocognitive

---

- ~ Subtle cognitive deficits
- ~ Unexplained mental retardation
- ~ Post-prandial loss-of-consciousness
- ~ Parkinson-like
- ~ Hepatic myelopathy
- ~ Pyramidal deficits

**Neurocognitive presentation most frequent in adults**

# Endocrine

---

- ~ Hyperinsulinemic hypoglycemia
- ~ Hyperandrogenism
- ~ Amenorrhea
- ~ Precocious puberty
- ~ Hypothyroidism

# Kidney

---

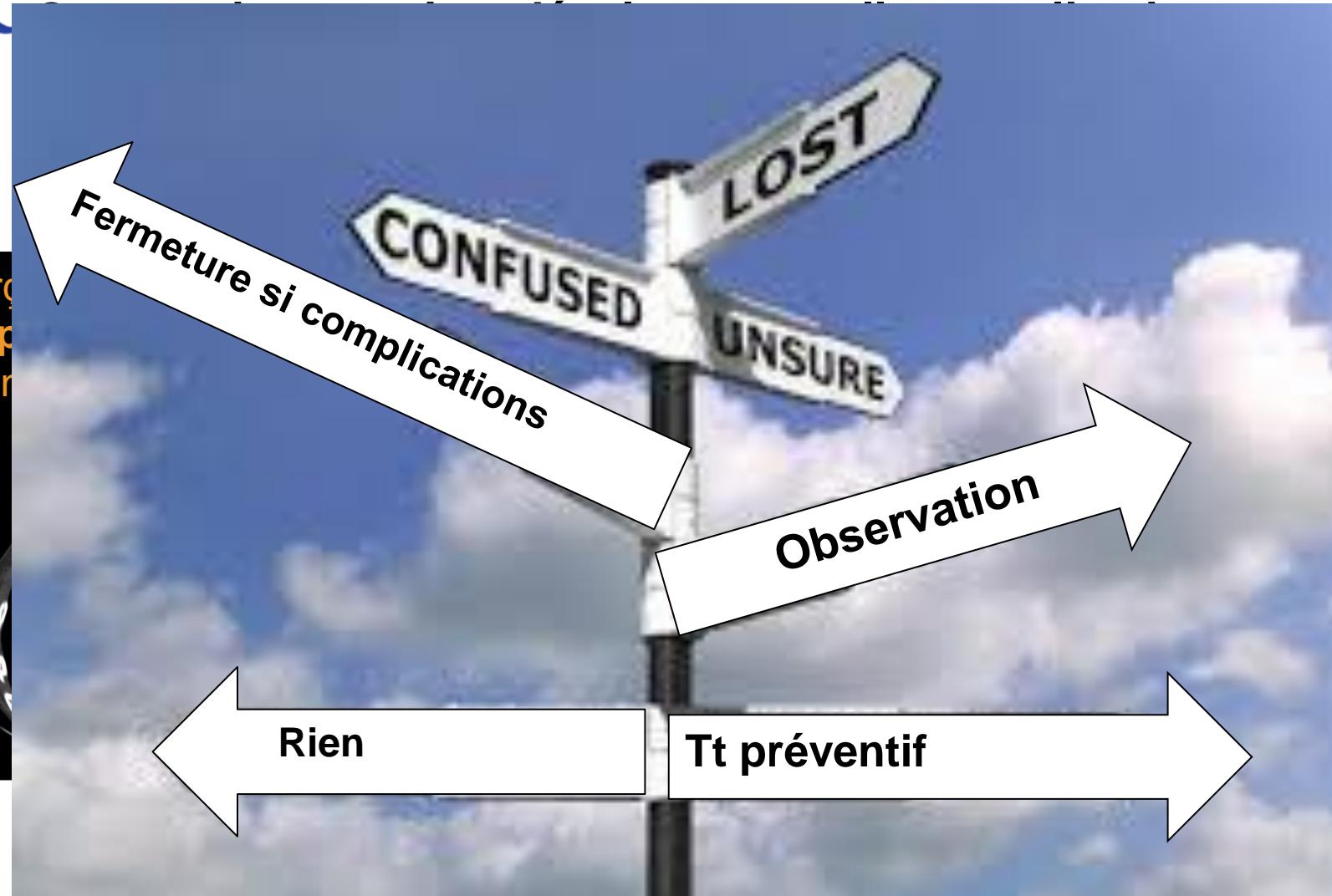
❖ Proteinuria

❖ Hematuria

# Existe-t'il des facteurs pronostiques des complications secondaires aux FPSC?

and

Gard  
resp  
shur



85 ans

# Recommended work-up

- Abdominal US + Doppler
- CT angiography → portal venogram

**Evaluate the shunt**

- Cardiac contrast echo
  - If pressures abnormal
    - Right heart catheterization
- Lung scan (HPS)
- Brain MRI & behavioral assessment
- Liver biopsy—underlying liver disease?
- Liver biochemistry, aFP, coagulation, bile acids, NH3
- Look for other malformations

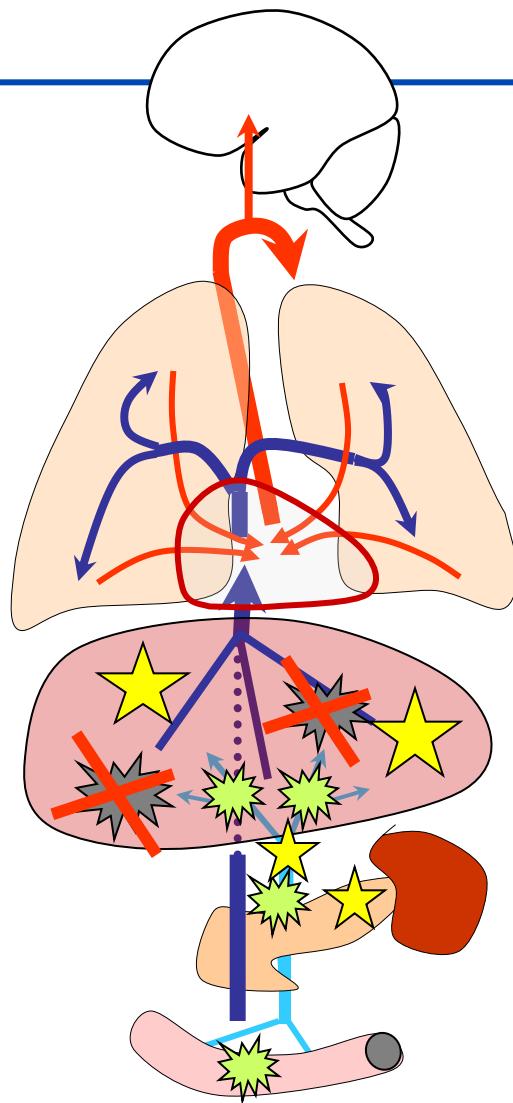
**Assess complications**

# Known associations-syndromic

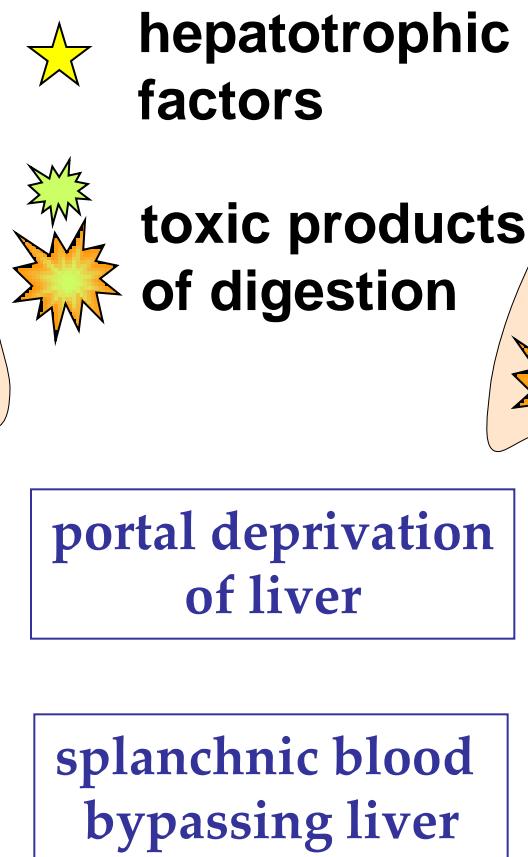
- Caroli's
- Goldhenhar
- Down's
- Turner
- Leopard
- Rendu-Osler
- Grazioli
- Noonan
- Cornelia de Lange
- Holt-Oram
- Costello
- Wolf-Hirschhorn
- Neurofibromatosis
- Adams-Oliver

No clear candidate genes or heritability  
patterns

# Physiological changes related to CPSS



Normal anatomy



CPSS

Courtesy of Pr F Gauthier

---

# Evaluation en imagerie des FPSC

Stéphanie FRANCHI-ABELLA  
Service de radiopédiatrie- Hôpital Bicêtre- Hôpitaux universitaires Paris-Sud

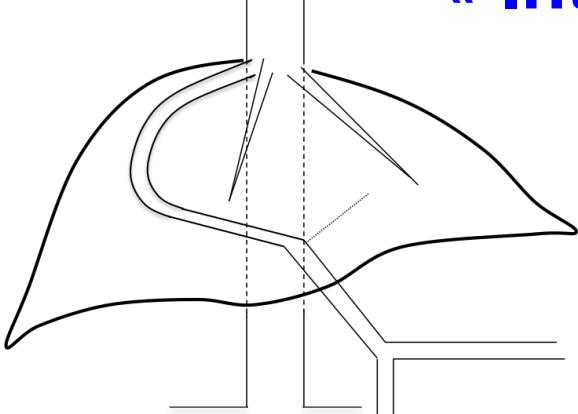
# Objectifs

---

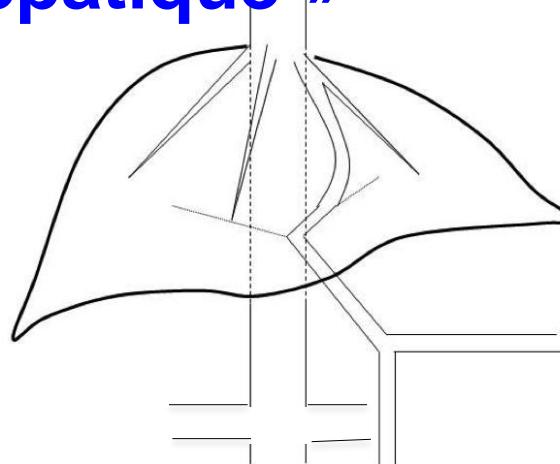
- ~ Formes anatomiques
- ~ Evolution dans la prise en charge des FPSC
- ~ Evaluation pré-thérapeutique
- ~ Techniques de fermeture

# Formes anatomiques de FPSC- classification de Bicêtre

## « Intrahépatique »



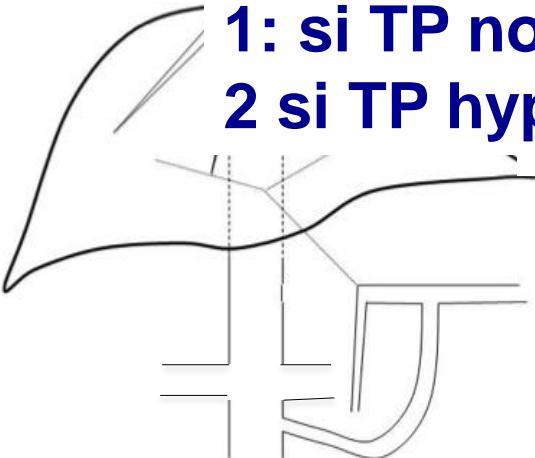
Porto-sus-hépatique



Ductus Venosus malformatif

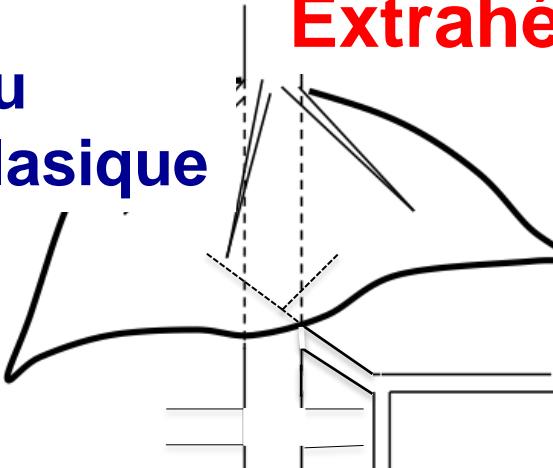
## Abernethy:

- 1: si TP non vu
- 2 si TP hypoplasique

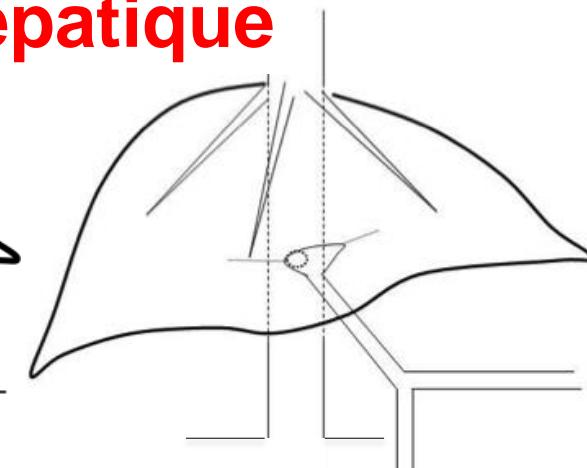


Porto-systémique  
Extra-Hépatique

## Extrahépatique

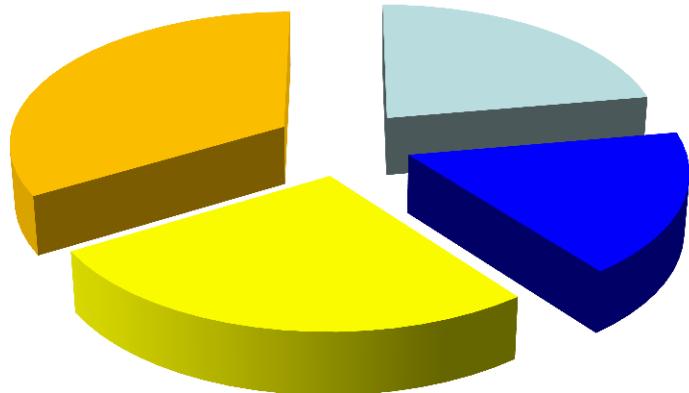


Porto-cave  
termino latérale



Porto-Cave  
latéro-latérale

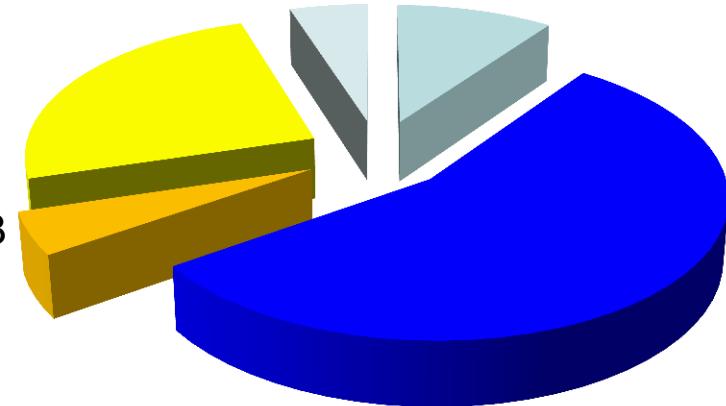
# Répartition des formes anatomiques



Revue, 265 pts

O Bernard, Seminars in Liver disease, 2012

- Ductus Venosus = type 2
- PortoHepatic= type 1
- PortoCaval = type 3
- Before Main Portal Vein type 4
- Complexe type 5



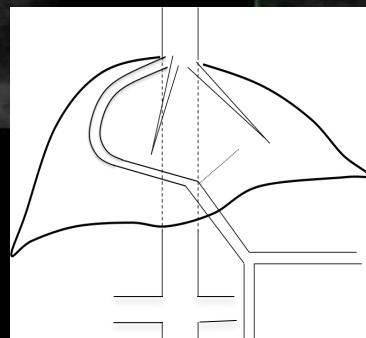
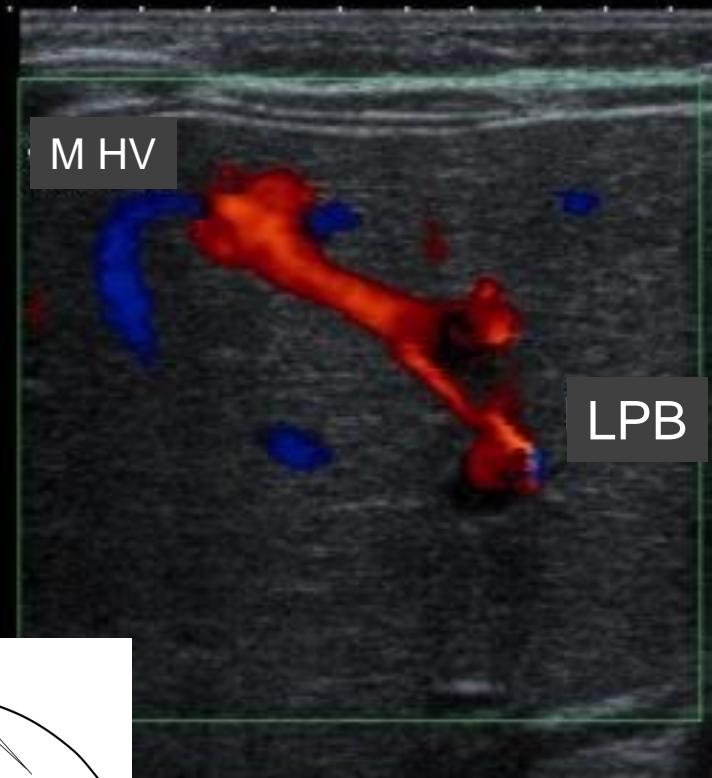
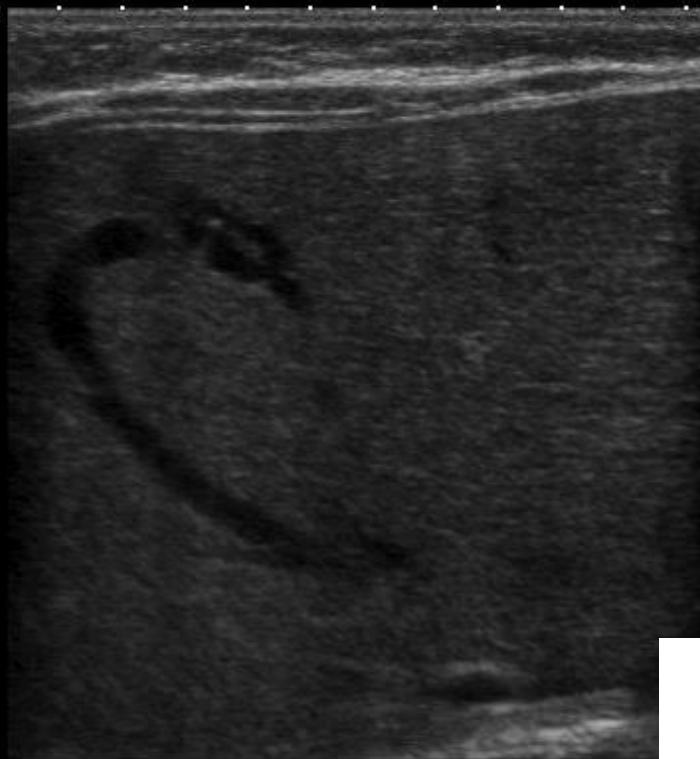
Série personnelle, 111 pts

Bicêtre, 2015

# Shunt porto-sus-hépatique

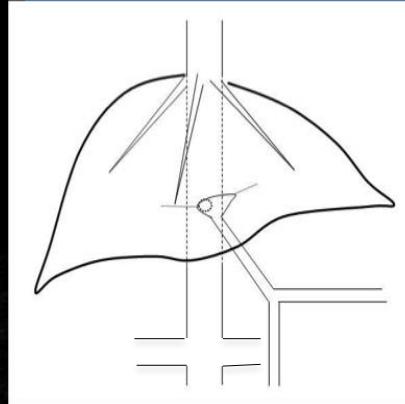


Diagnostic anténatal



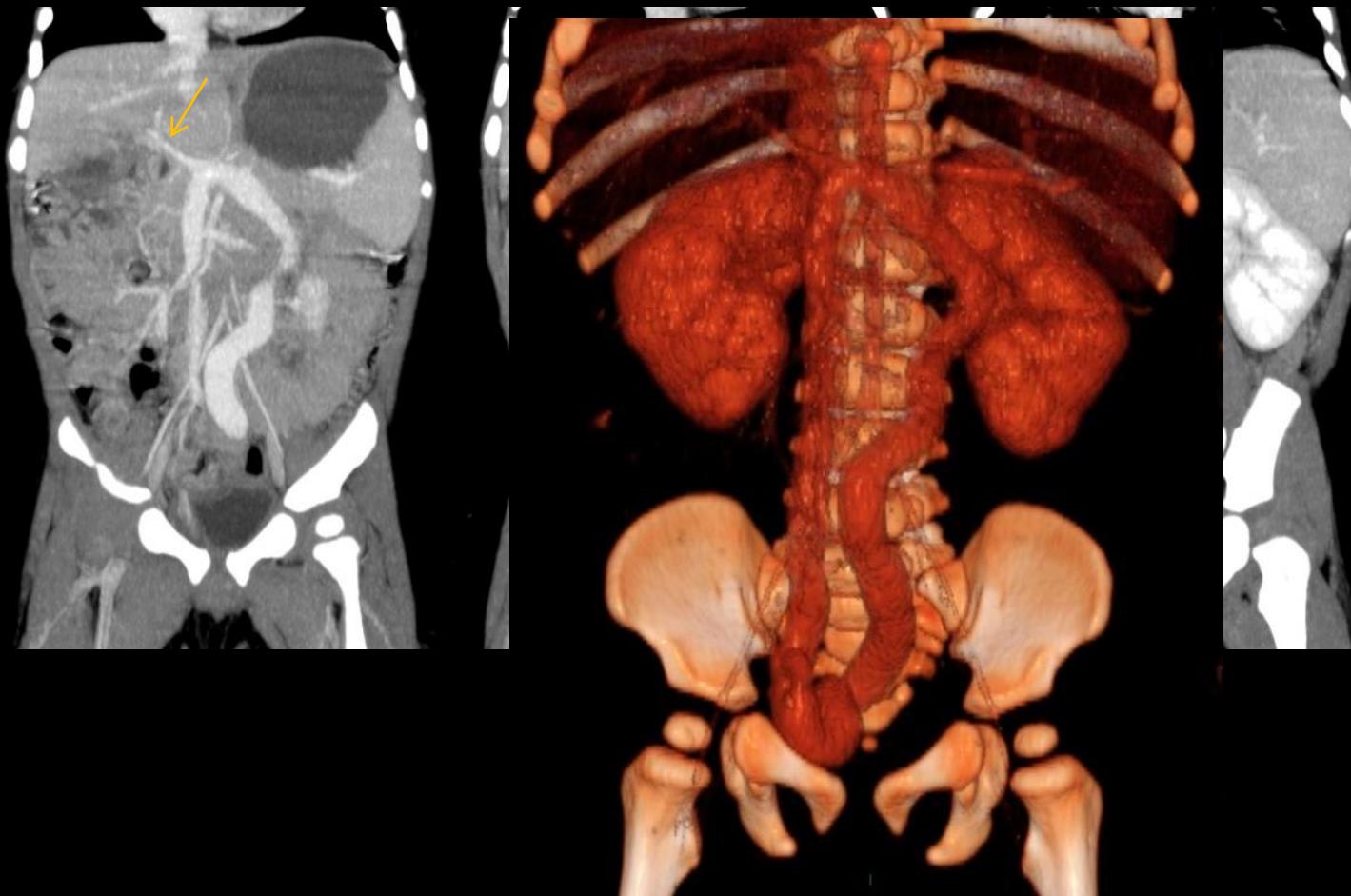
# Fistule porto-cave latéro-latérale

Difficultés scolaires

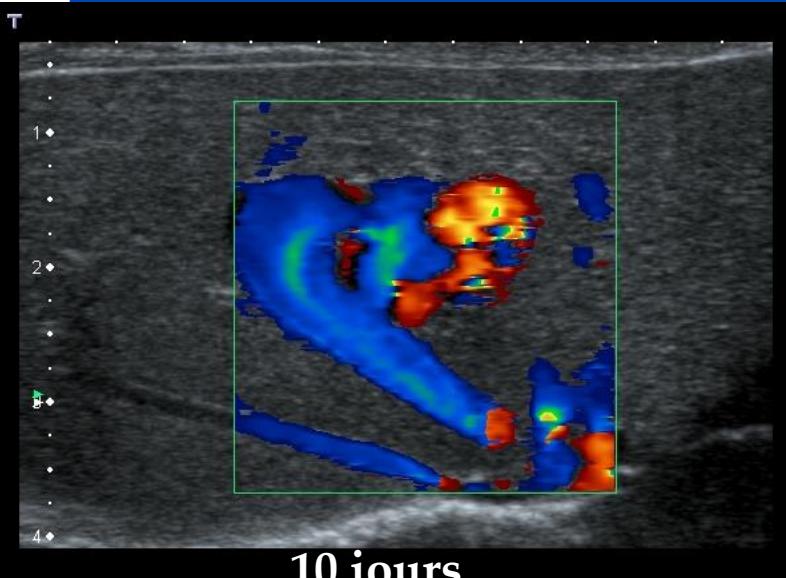


# Fistule entre la veine splénique et la veine iliaque interne droite, latéro-latérale

## Gros membre inférieur droit



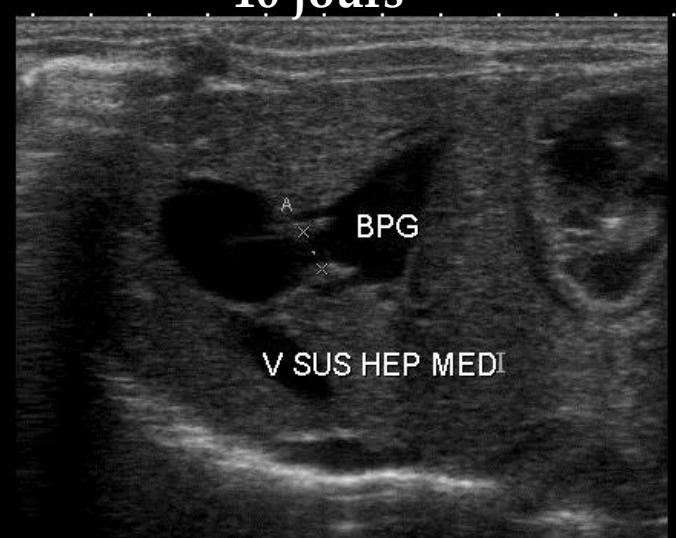
# Faut-il fermer tous les shunts et quand?



10 jours



2 ans



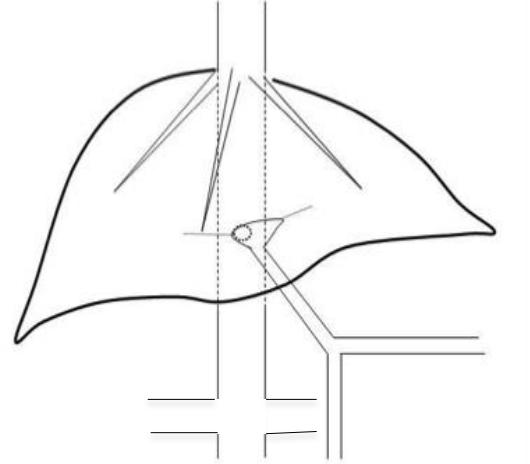
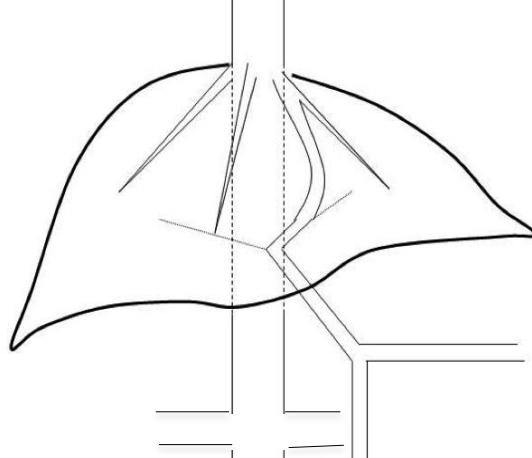
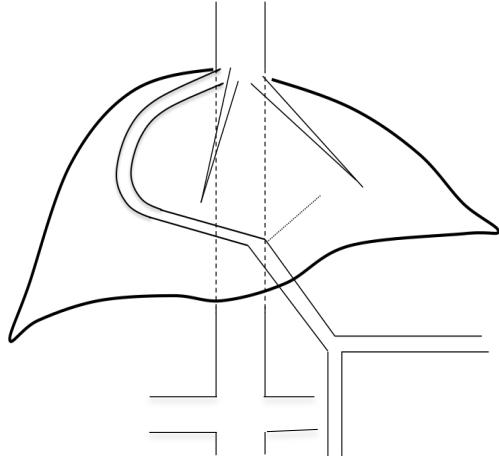
**Non**

**Fermeture spontanée jusqu'à 2 ans  
des fistules porto-sus-hépatiques  
= surveillance**

**Si la fistule porto-sus-hépatique persiste  
Ou s'il s'agit d'une autre forme  
anatomique = fermeture à discuter**

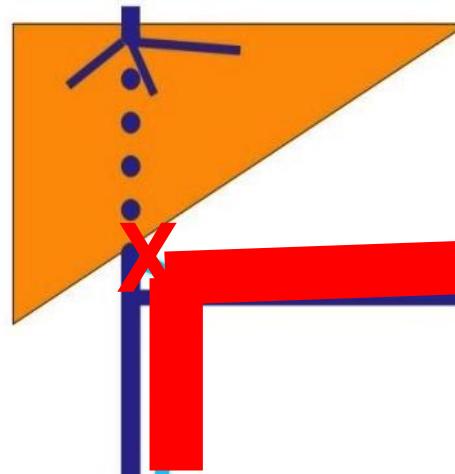
# Fermeture des FPSC

## Quand le tronc porte est visible...



Développement attendu du TP et des branches portes intrahépatiques après fermeture

Risque théorique d'HTP massive dans la littérature:

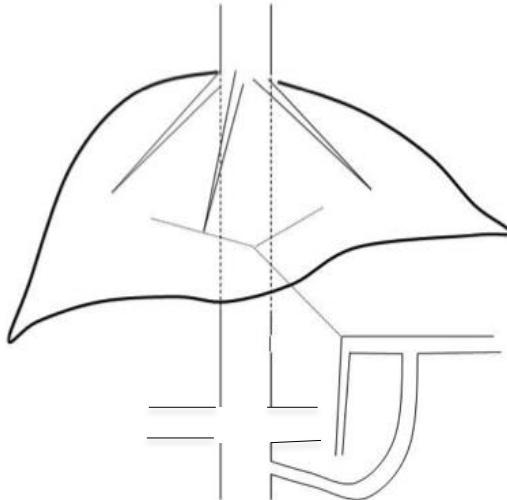


Abstention  
TH

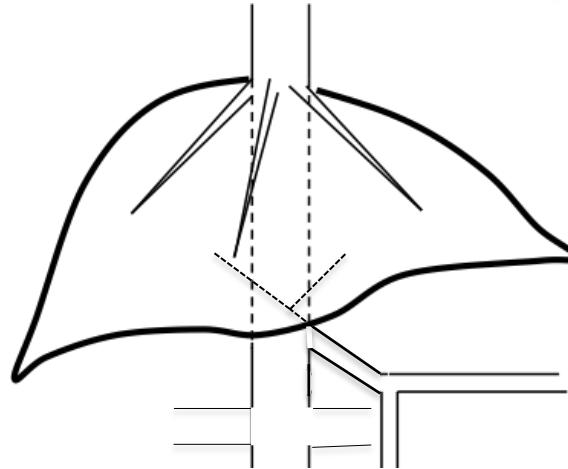
Que faire?

# Evaluation avant fermeture

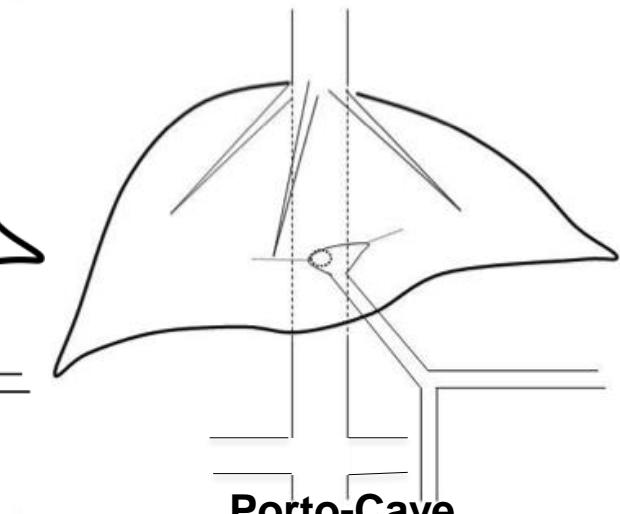
## Intérêt du test d'occlusion



**Porto-systémique  
Extra-Hépatique**



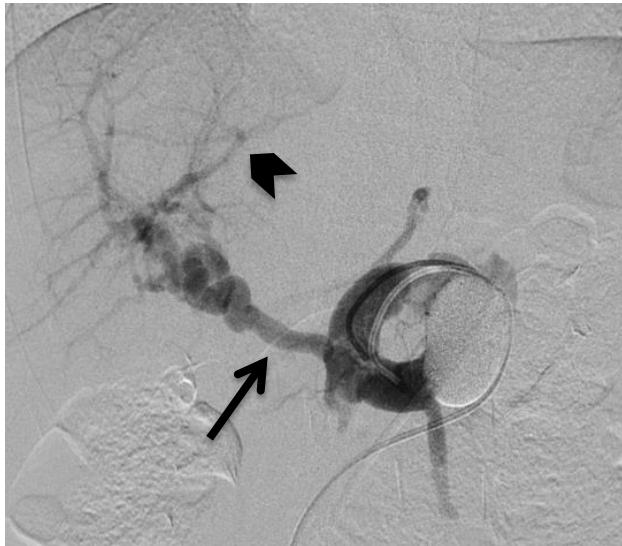
**Porto-cave  
termino latérale**



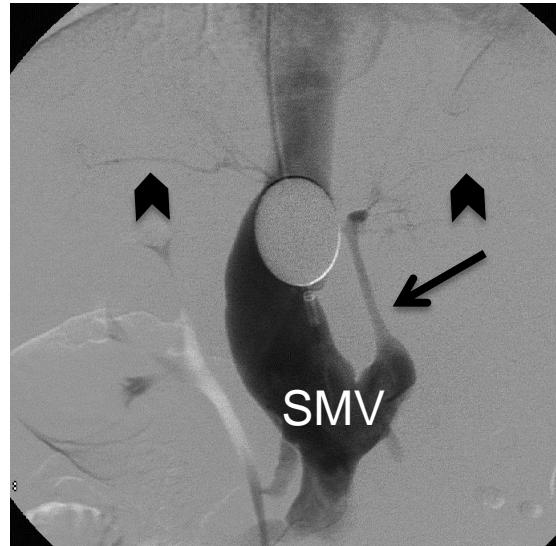
**Porto-Cave  
latéro-latérale**

- Pour préciser la présence d'une veine porte hypoplasique et/ou ectopique et l'aspect des branches portes intrahépatiques
- Pour mesurer la pression portale= choix de fermeture en 1 ou 2 temps

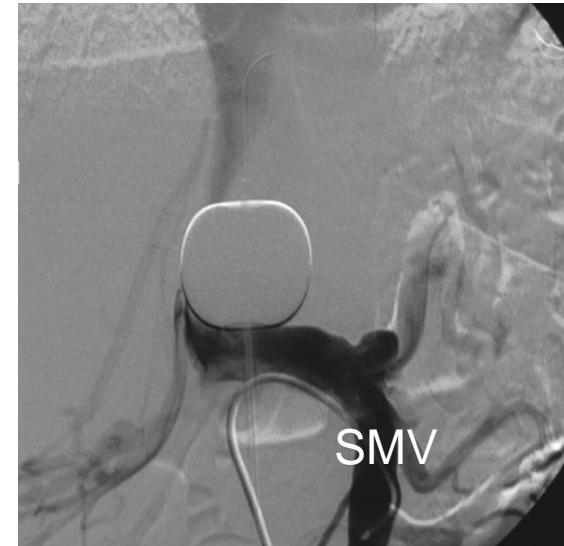
# Classification du système porte intrahépatique lors du test d'occlusion proposée par Kanazawa H. J Pediatr Surg 2015



Hypoplasie faible

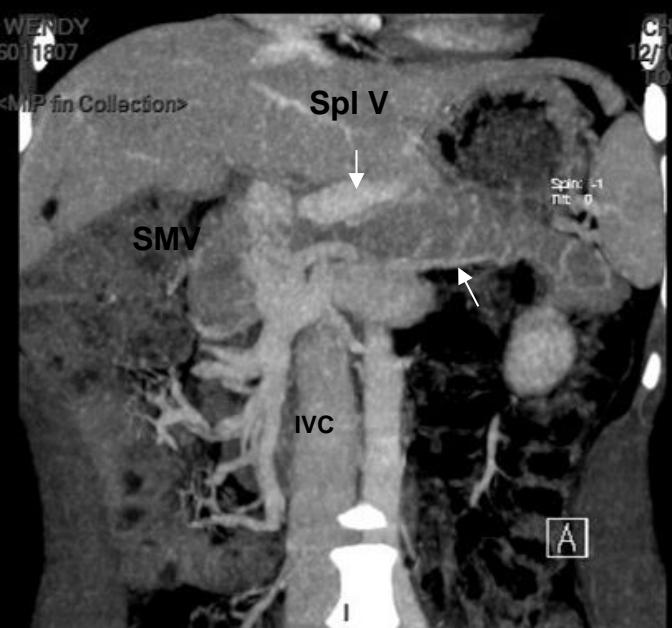
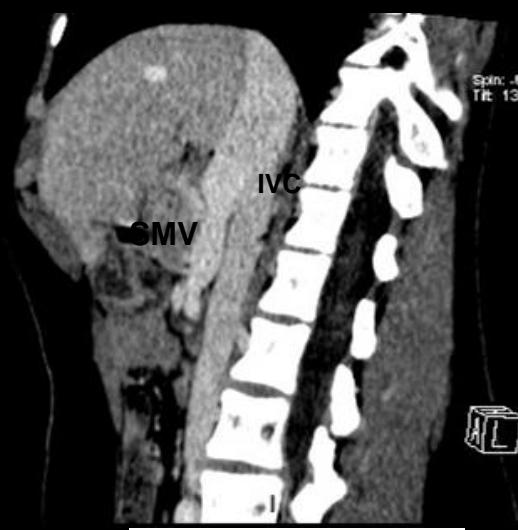
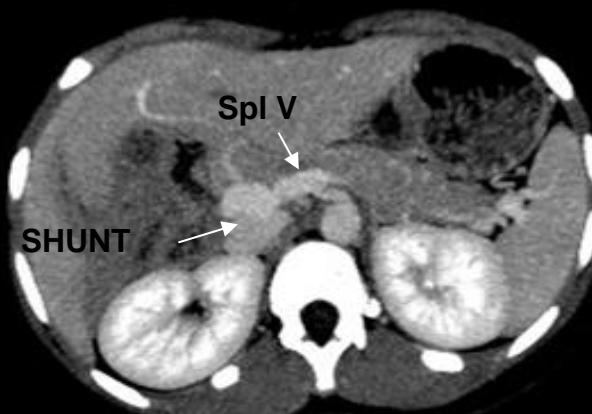
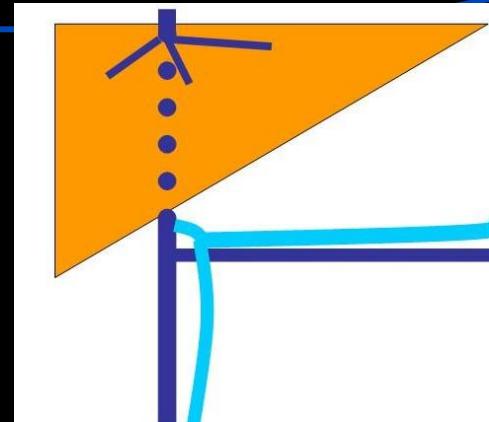
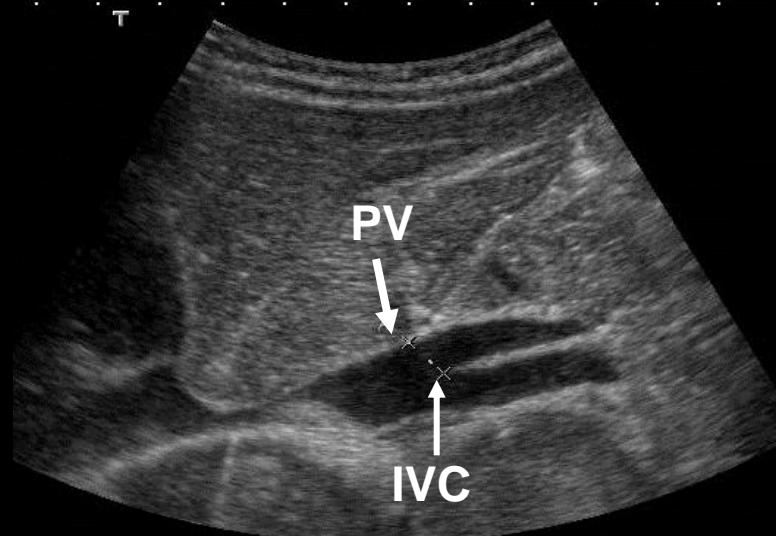


Hypoplasie modérée



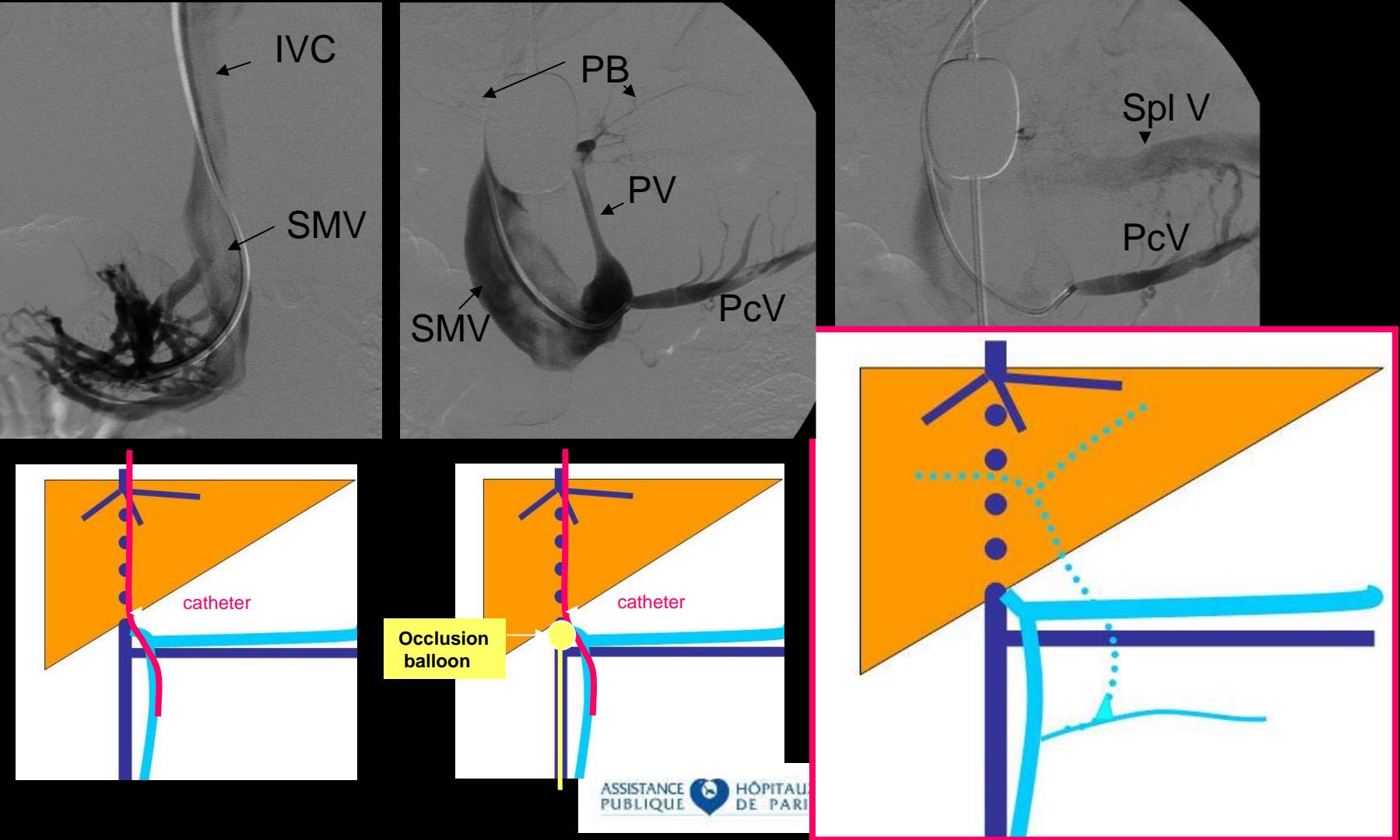
Pas de flux hépatopète

# DAN, Encéphalopathie hépatique Diagnostique de fistule porto-cave termino-latérale

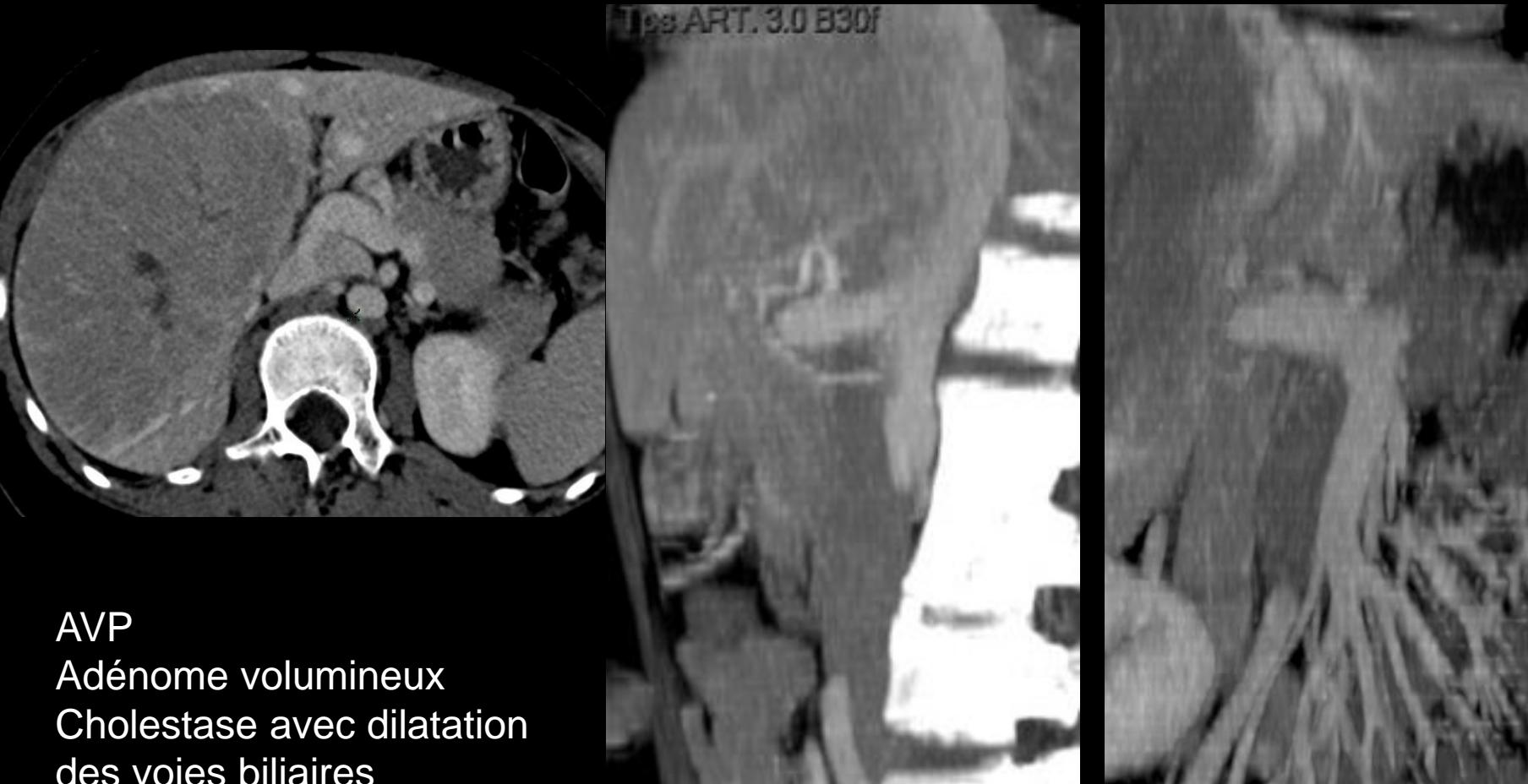


# Fistule porto-cave latéro-latérale

## Angiographie avec test d'occlusion

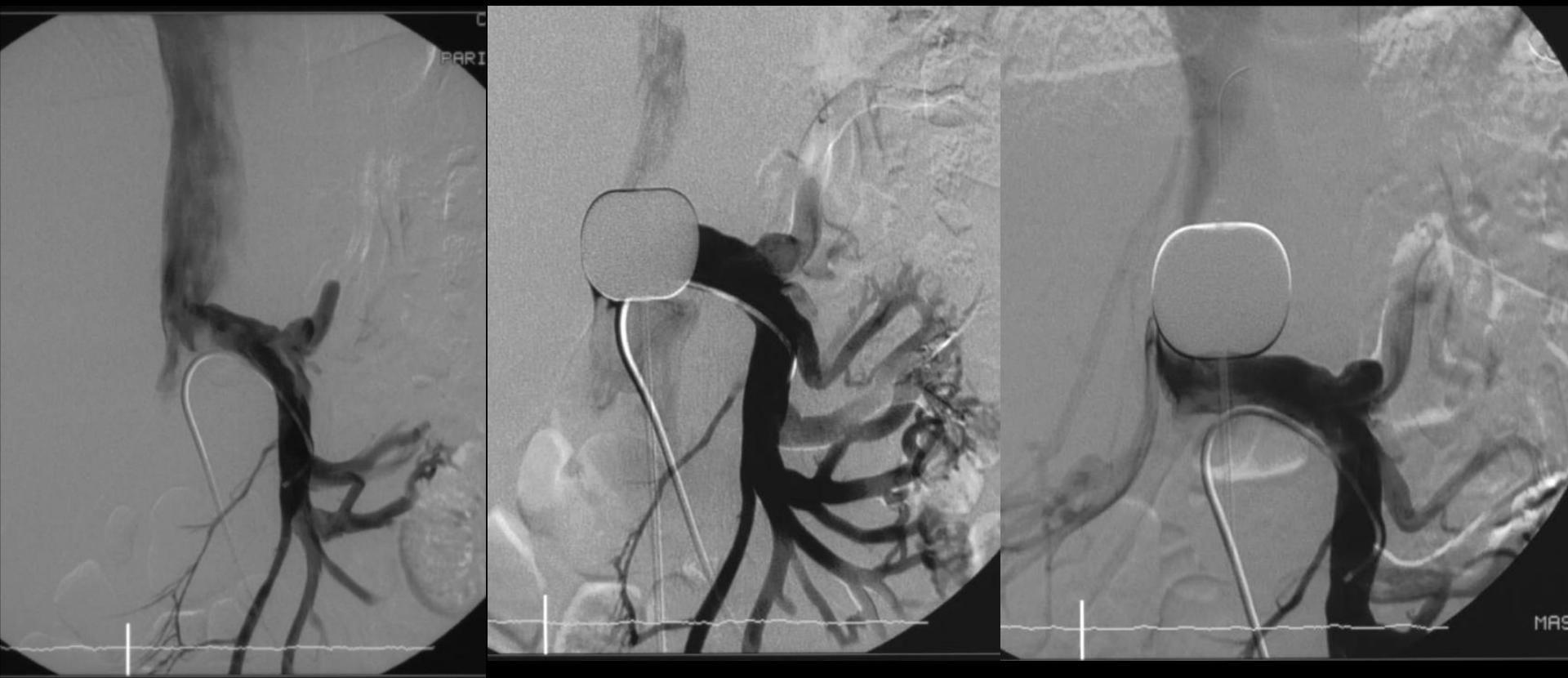


# Fermeture d' une fistule porto-cave termino-latérale (Abernethy 1)



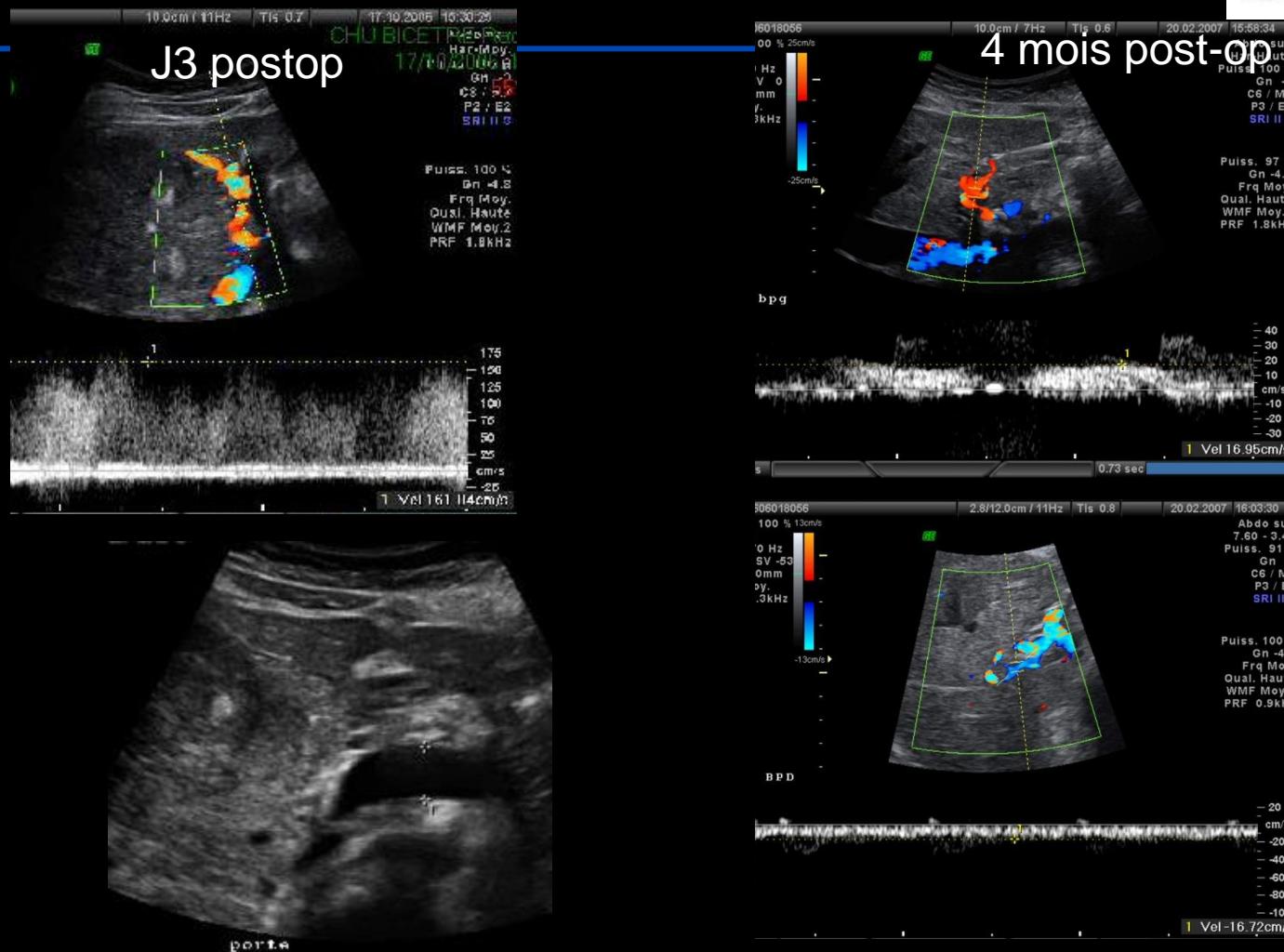
AVP  
Adénome volumineux  
Cholestase avec dilatation  
des voies biliaires

# Evaluation préthérapeutique Angiographie test d'occlusion



Fistule porto cave termino-latérale pré-hépatique. Pas de TP et de SPIH visibles à l'occlusion.  
Pression portale à l'occlusion 20 mmHg.  
Décision de fermeture chirurgicale **en deux temps**

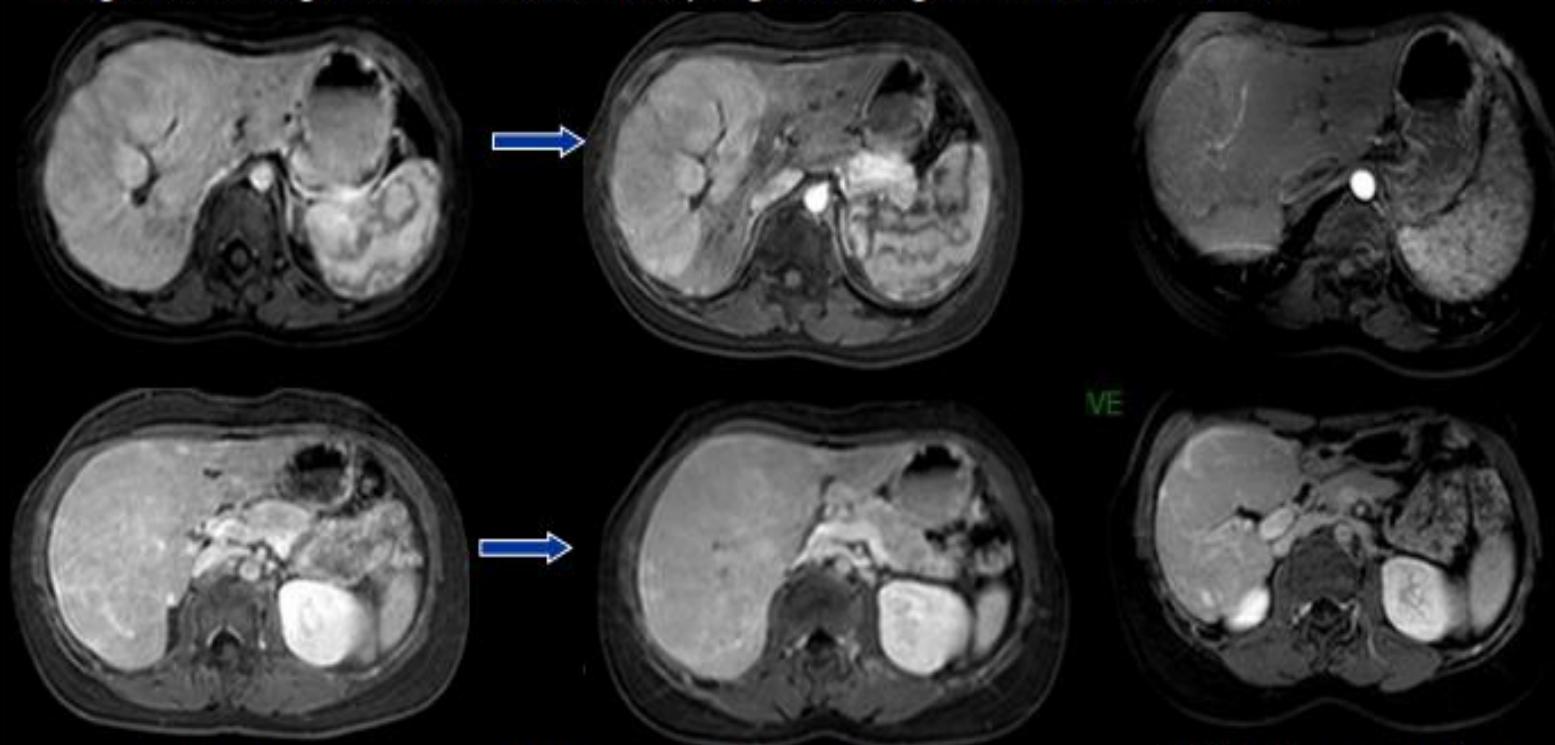
# Suivi post-opératoire ( cerclage)



Apparition d'un flux porte intrahépatique hépatopète



A tortuous vein restores hepatopetal portal flow in the portal branches that developed  
Progressive regression of the HCA, progressive growth of normal liver



April 2006

2007 : 2 m, post-binding

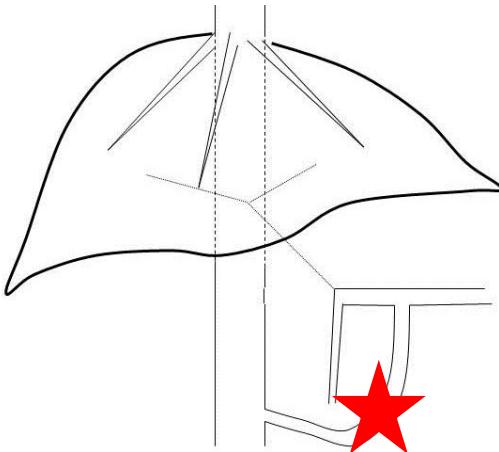
2012: 5 y. post closure

# Choix de la technique d'occlusion

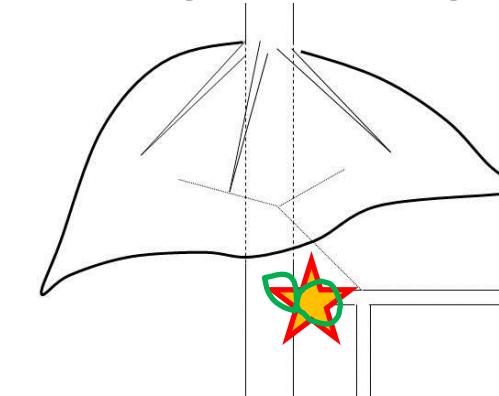
## O Fermeture par voie radiologie interventionnelle si possible

**Anticoagulation +++**

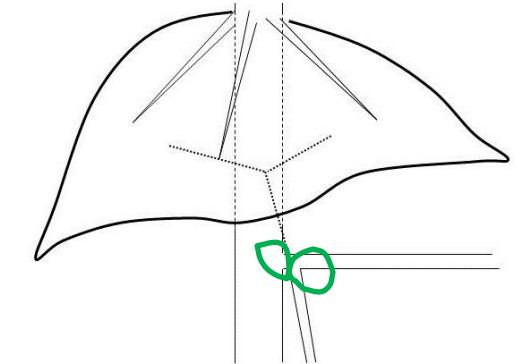
Assez long et pas trop large  
= RI



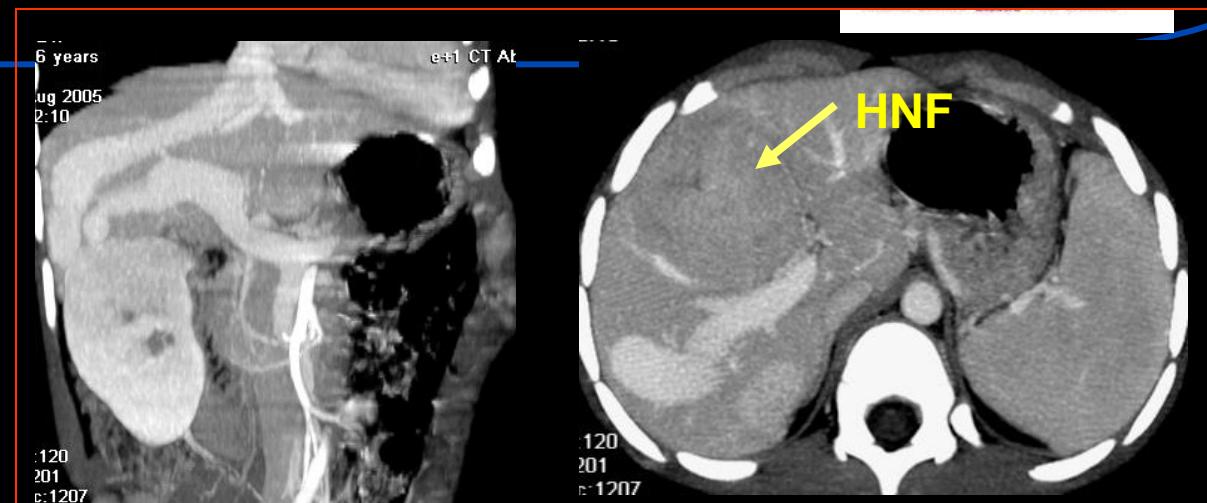
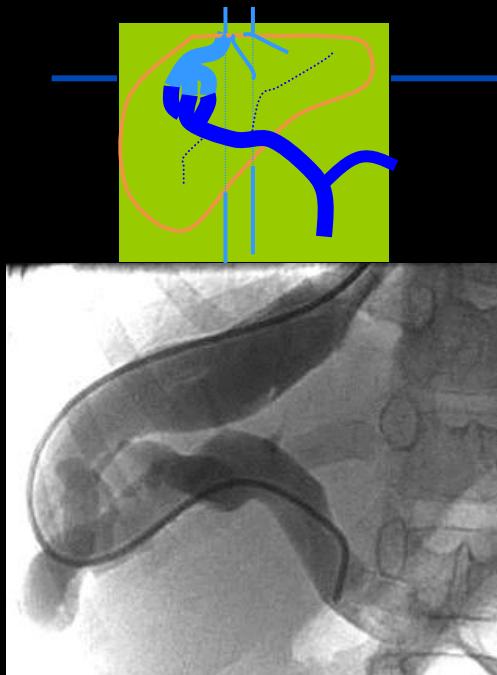
Radiologie ou chirurgie...



Trop court pour mettre un dispositif  
= chirurgical



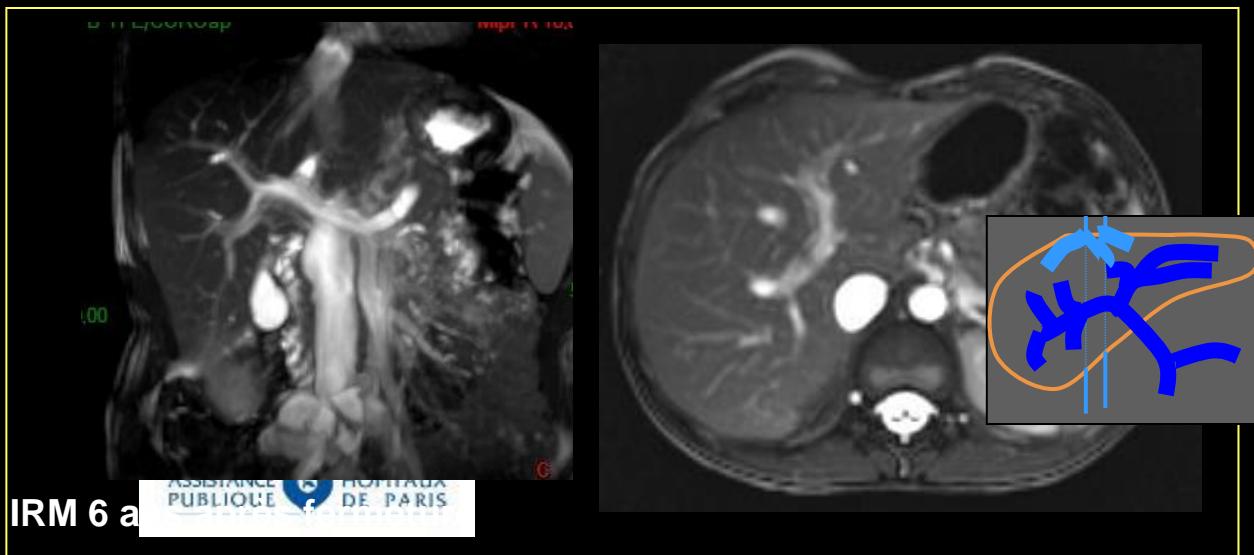
# Fistules porto-sus-hépatiques multiples compliquées d' HNF



Avant fermeture



Angiographie et occlusion des fistules par des plugs



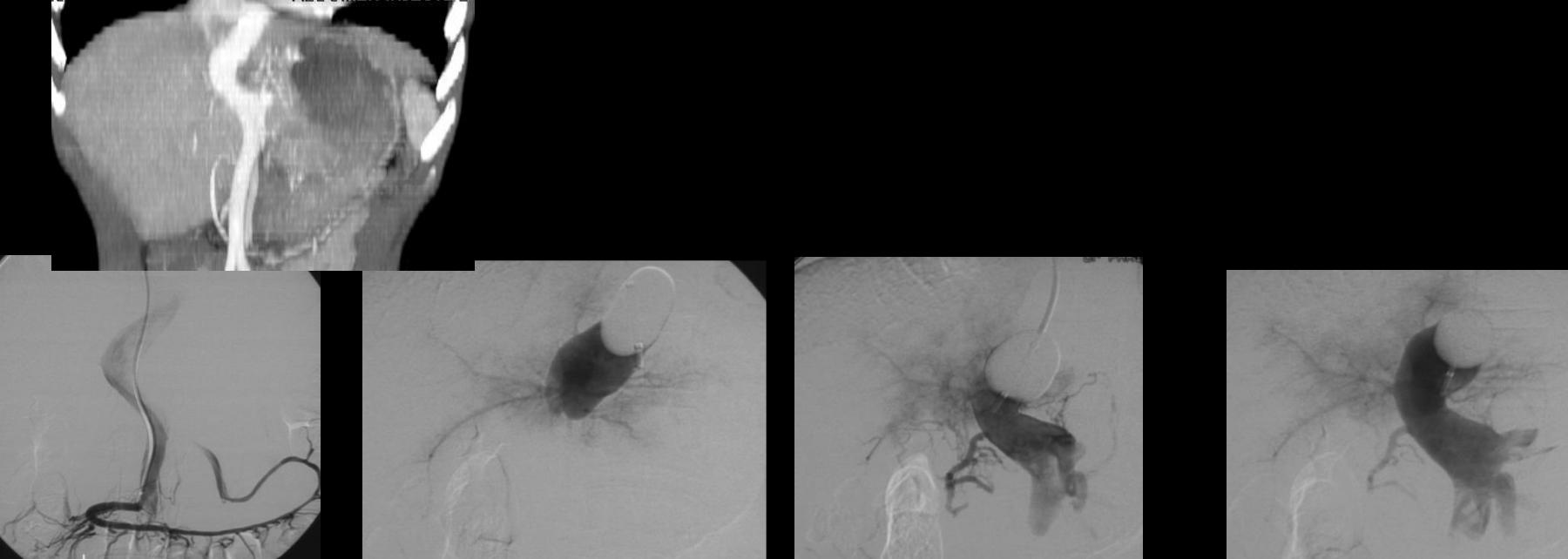
**Garçon de 15 ans**

**Découverte fortuite d'un TP à 50% avant plastie des oreilles**

**ATCD d'insuffisance cardiaque non étiquetée traitée de la naissance à 3 ans**

24

ABDOMEN INJECTÉ 2



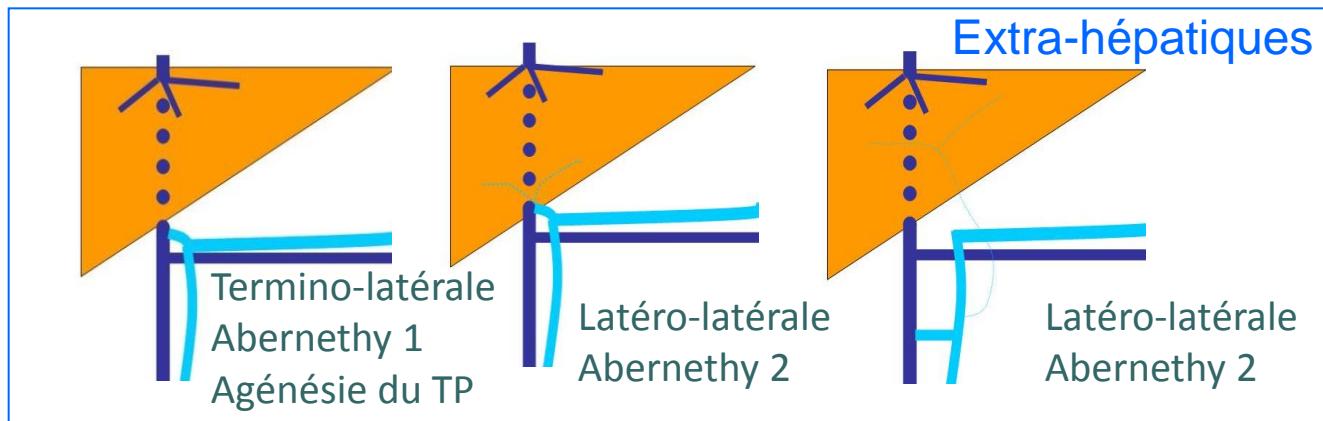
Fermeture Amplatzer  
07.2004



# Existe-t'il des cas où la fermeture néonatale est souhaitable?

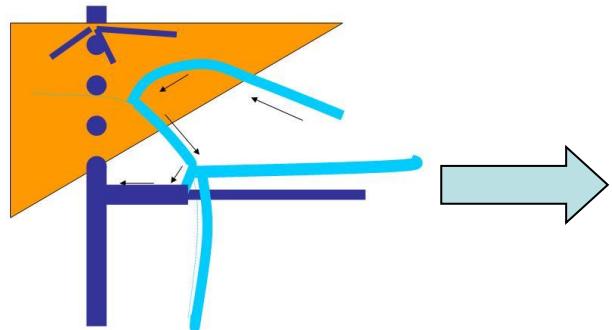
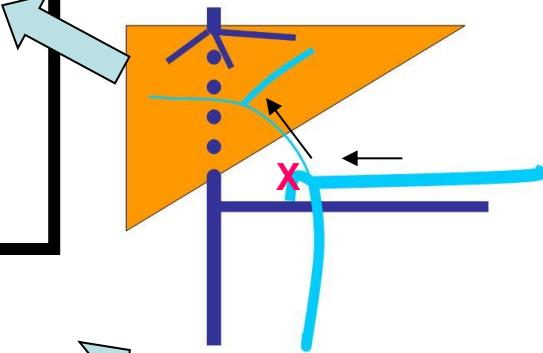
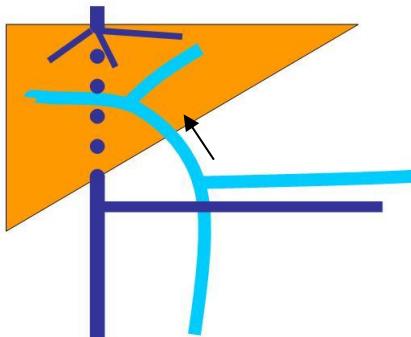
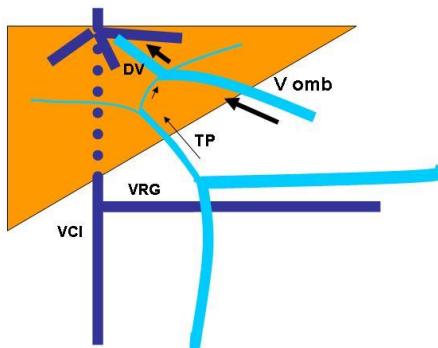
Oui!

**En cas de fistule en amont de la bifurcation portale**

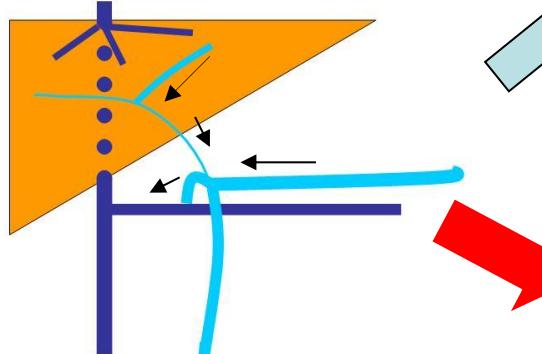


# Histoire naturelle des FPSH extra-hépatiques

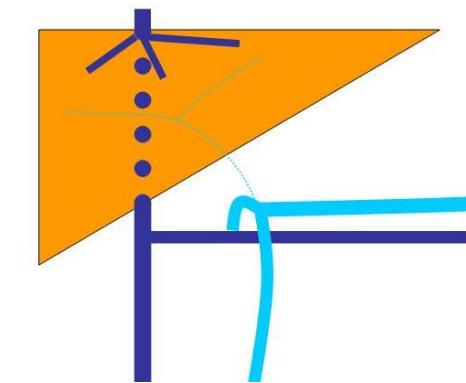
## Aspect normal pré et post-natal



Pré-natal



Post-natal



Sans traitement

# Le point de vue du chirurgien

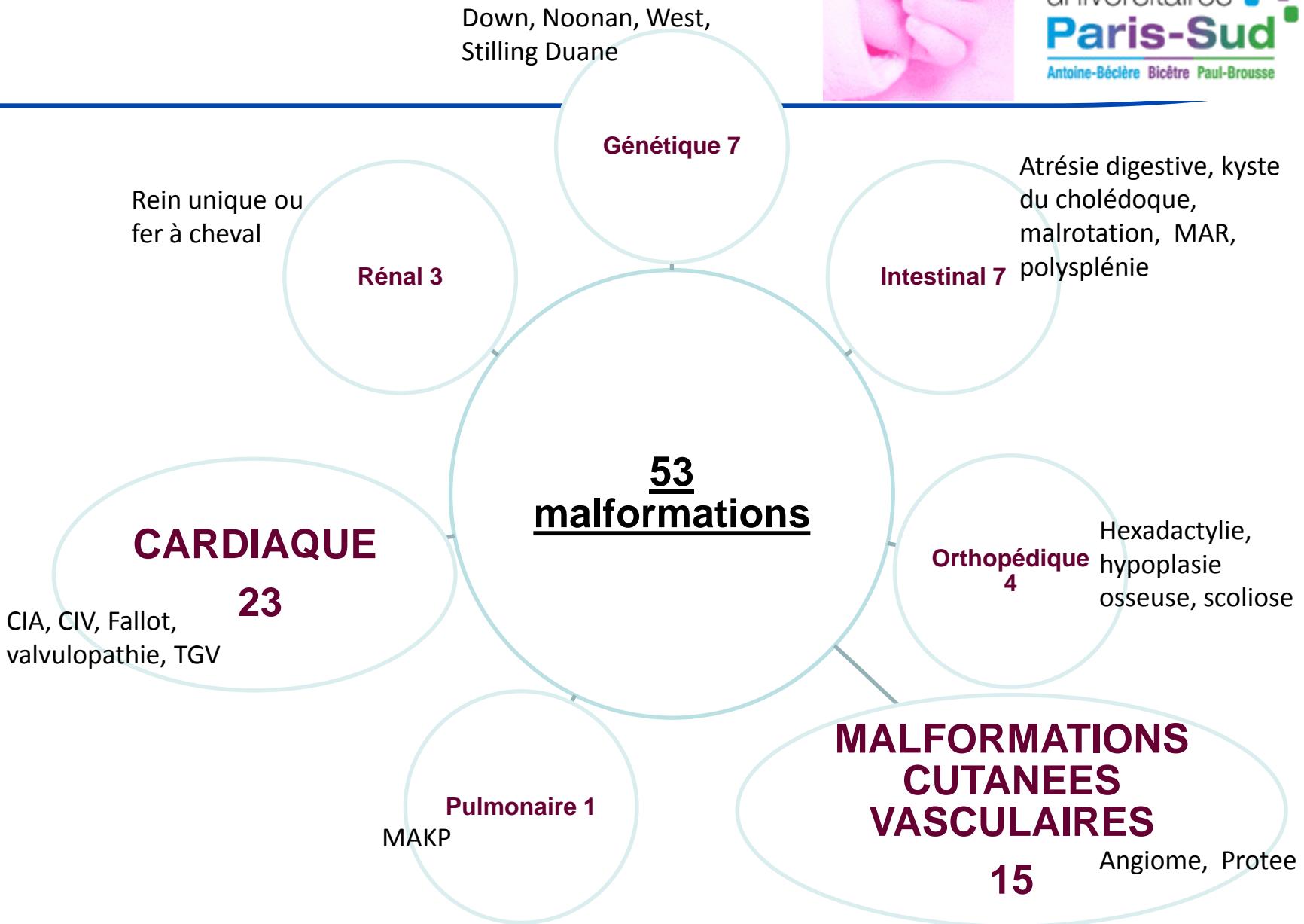
- Indication
- Evaluation
  - Le terrain
  - Les complications
  - L'anatomie
- L'opération
- Les suites



# Évaluation: Le terrain



100 pts



# RESULTATS



Hôpitaux  
universitaires  
**Paris-Sud**

	<b>malformations congénitales (n=53)</b>	<b>isolées (n=48)</b>	
fermeture spontanée	17	13	
fermeture complète	23	29	
-chirurgicale	12	17	
-radiologique	11	12	
fistule résiduelle	3	1	
en attente de fermeture	10	5	
total non fermée	13(25%)	6(12,5%)	<b>P=0,06</b>

# RESULTATS



	<b>CMCPS (n=53)</b>	<b>NoCMCPS (n=48)</b>
Complications du geste	2 migrations de plug 1 hémopéritoine 1 effraction vasculaire	1 migration de plug 1 hémopéritoine
Mortalité	1 hépatocarcinome métastatique	0
Transplantation hépatique	1	0

**P=0.16**

# Évaluation complications de la fistule

Respiratoire

Sd Hépatopulmonnaire

VNI post opératoire



Cardiaque

HTAP

Monitorage veineux central (Swan Gantz) et artériel

Malfo associée: CIA/CIV

Embolie gazeuse

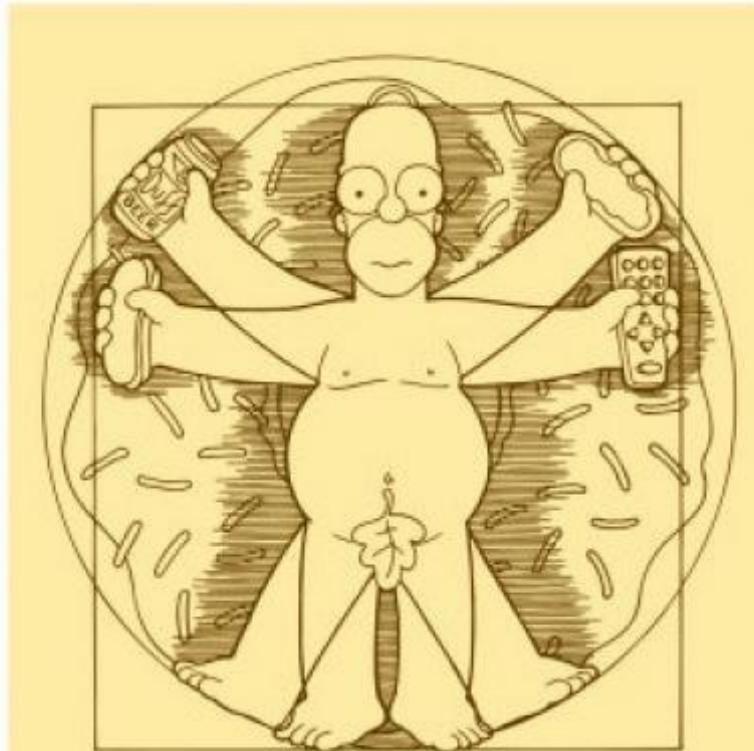
Hépatique:

Maladie hépatique sous jacente

Rôle de la biopsie?

Nodules  $\beta$ -cat mutés: Resection vs. surveillance vs.  
transplantation?

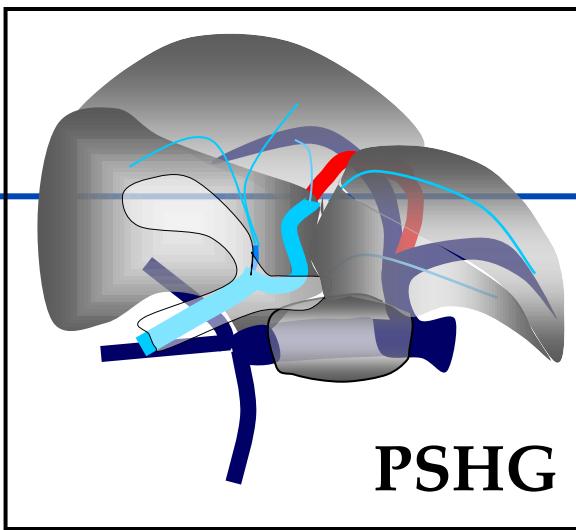
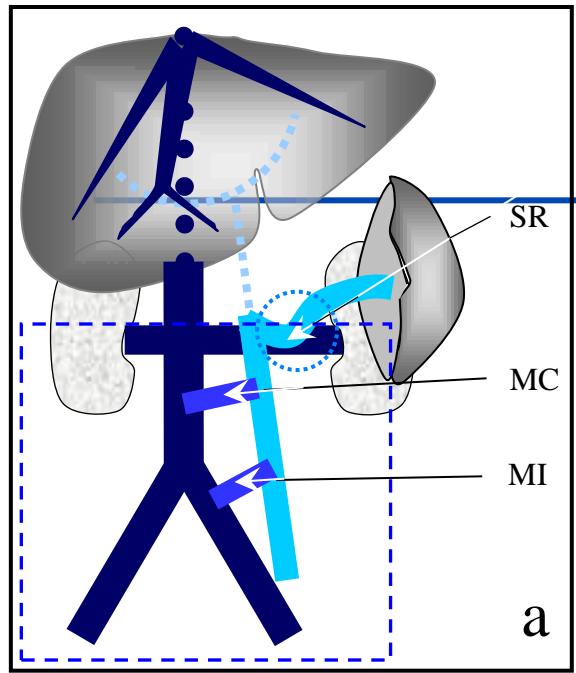
# Evaluation: L'anatomie?



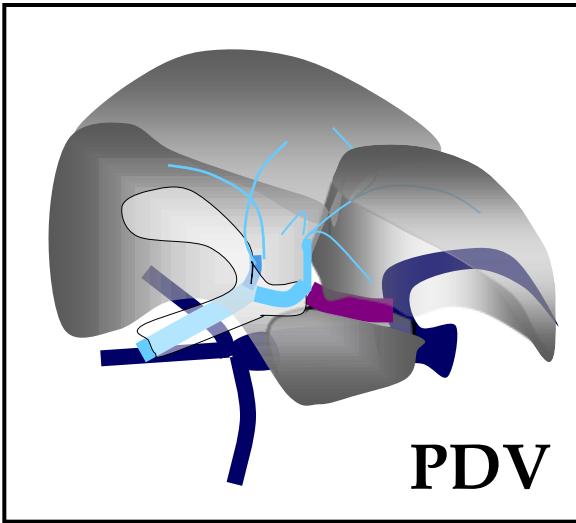
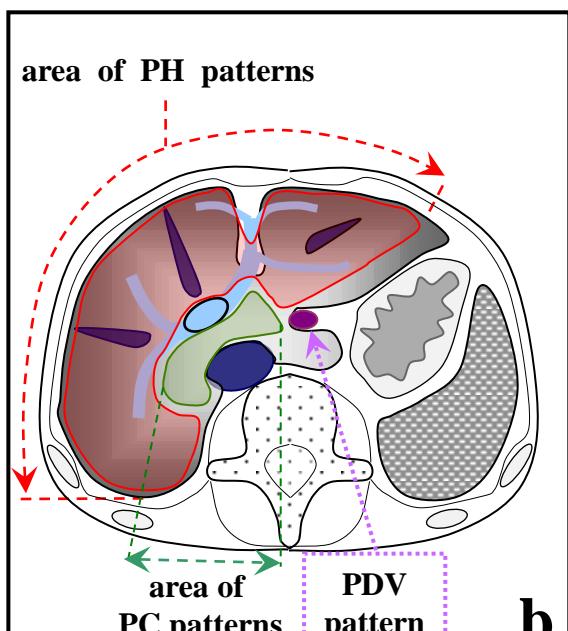
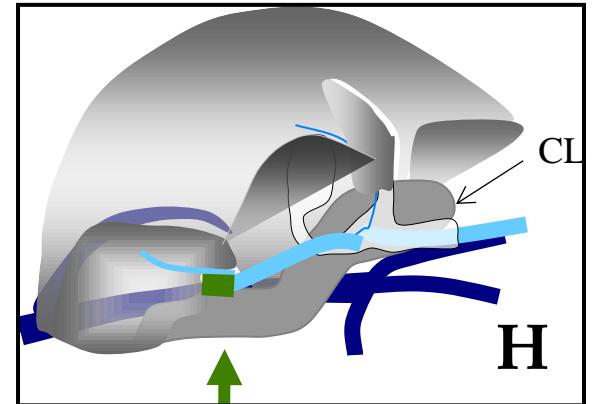
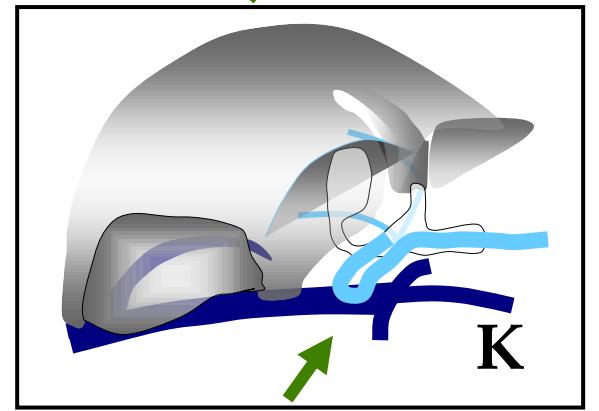
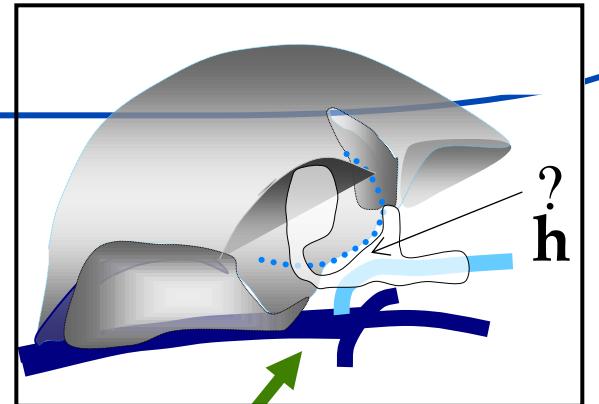
HOMER-HOMO NEANDERTHALIS

Mmm...Business Deal-Mmm...Crumbled-up Cookie Things  
Fresh Mmm...Mmm fresh-Mmm...Purple  
Mmm...Unprocessed Fish Sticks-Mmm...~~Big~~ Dollars

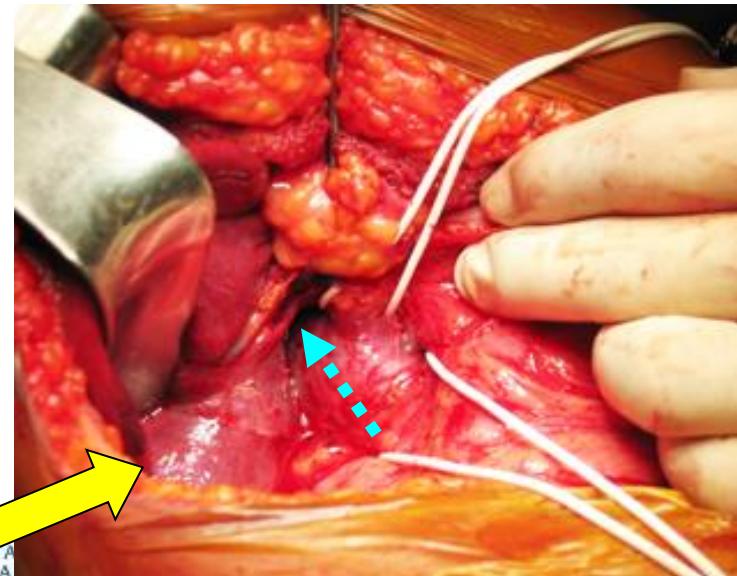
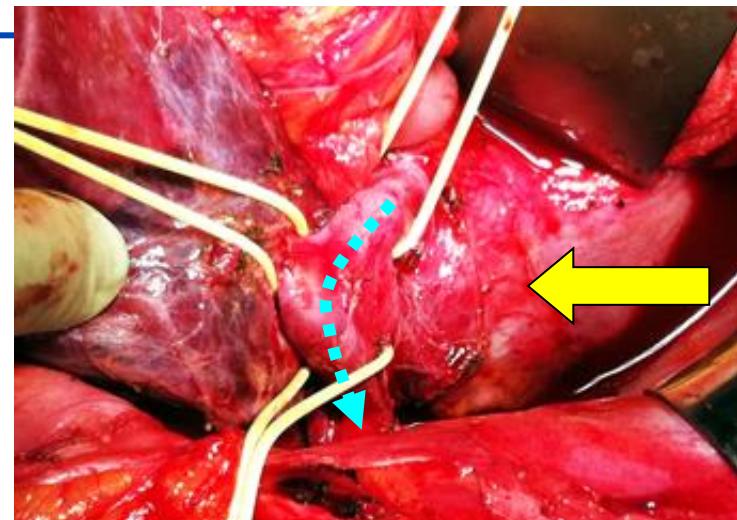
# Gauches



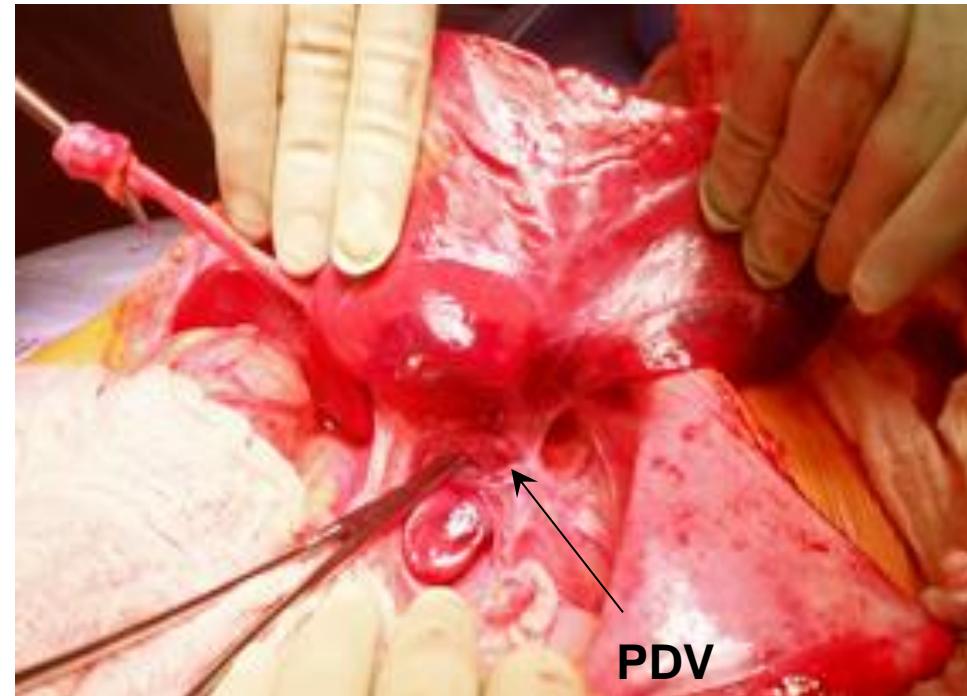
# Droites



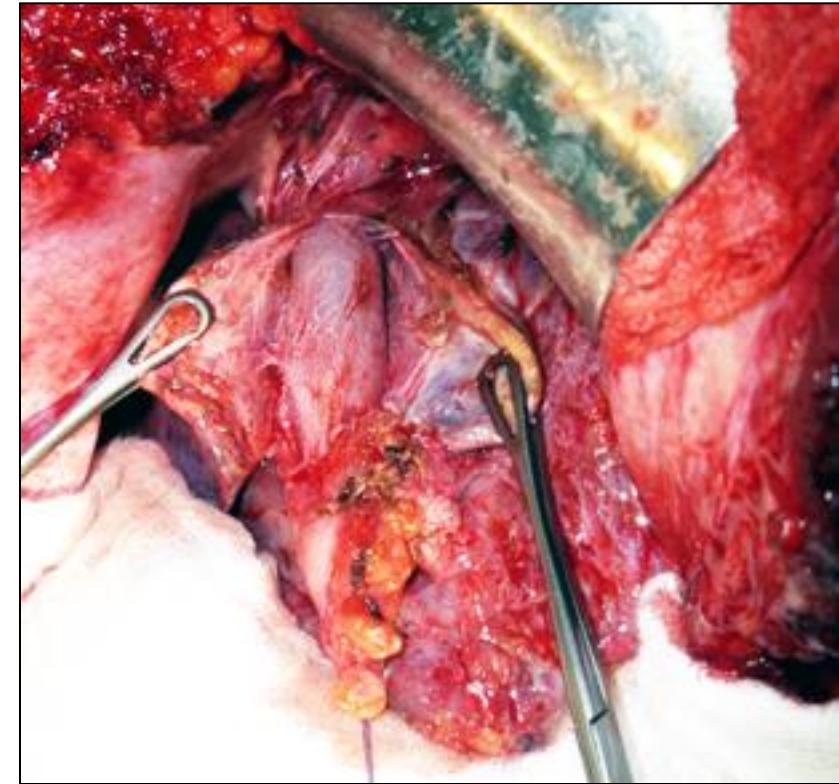
# FPC droites



# FPC gauches

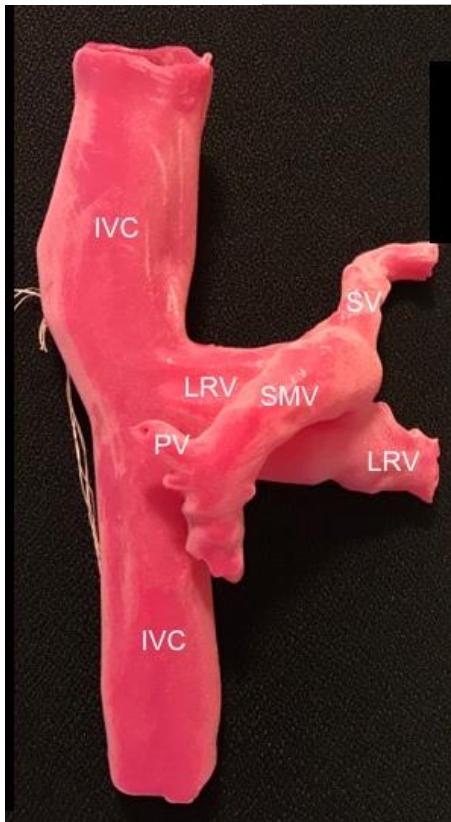


Persistent  
ductus venosus



FPC courant sur lobe gauche

# Les atypiques



# Fermeture chirurgicale

## 3 objectifs:

1) Epargner le système porte intra hépatique

→ Lier la partie cave du shunt

2) Prévenir la thrombose du système porte

→ anticoagulant

→ Doppler post op et adaptation de l'anticoag

3) Eviter la congestion intestinale

→ test de clampage

→ monitorage de pression portale

→ observation de l'intestin

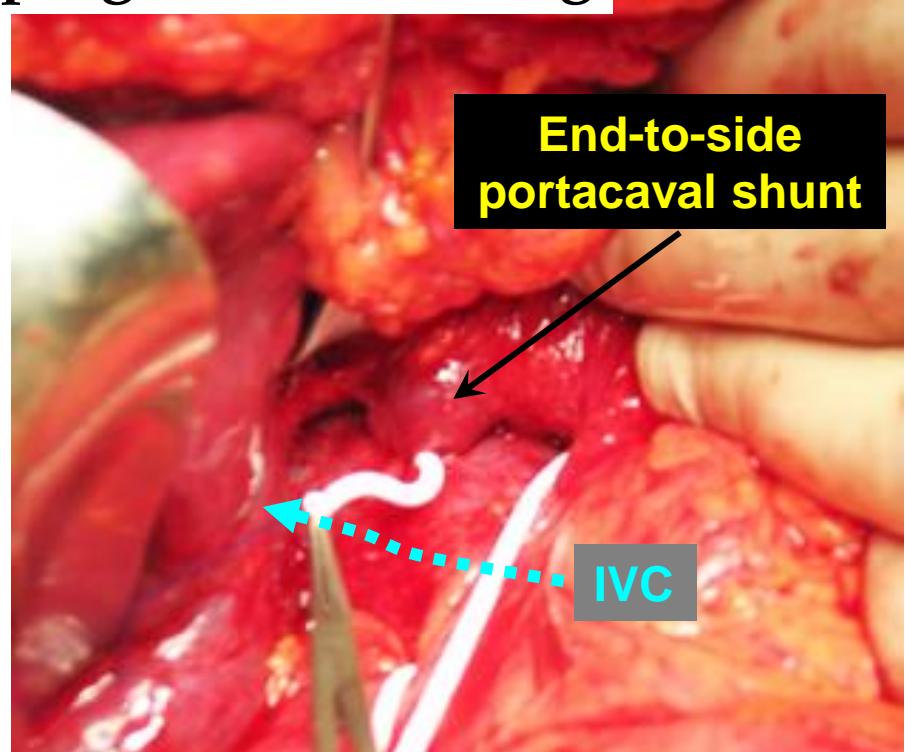
# Stratégie chirurgicale

---

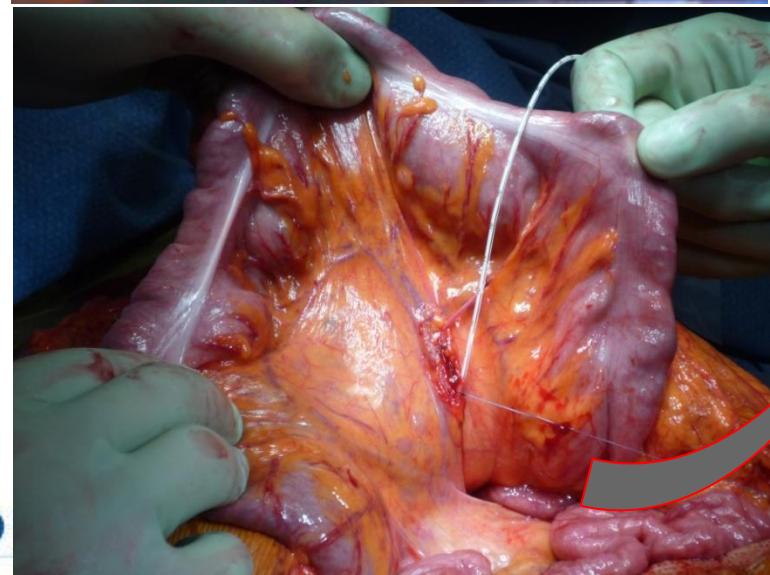
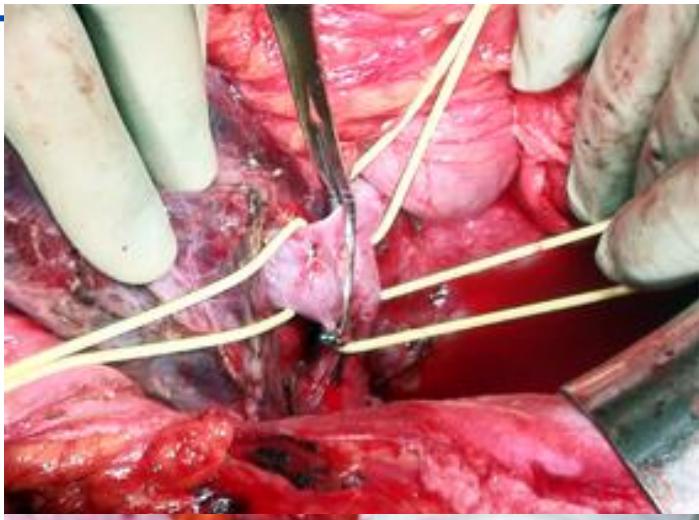
- ~ Identification de la fistule
- ~ Parfois sécuriser
  - Pringle
  - Préparation d'exclusion vasculaire
  - Risque embolie gazeuse
  - Monitorage anesth+++
- ~ Faire le tour (ouf!)
- ~ Monitorer les pressions portales

# From approach to action

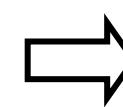
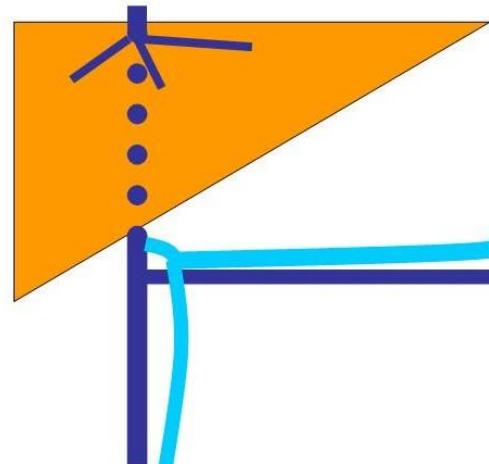
Portacaval CPS with  
poor tolerance to clamping test → banding



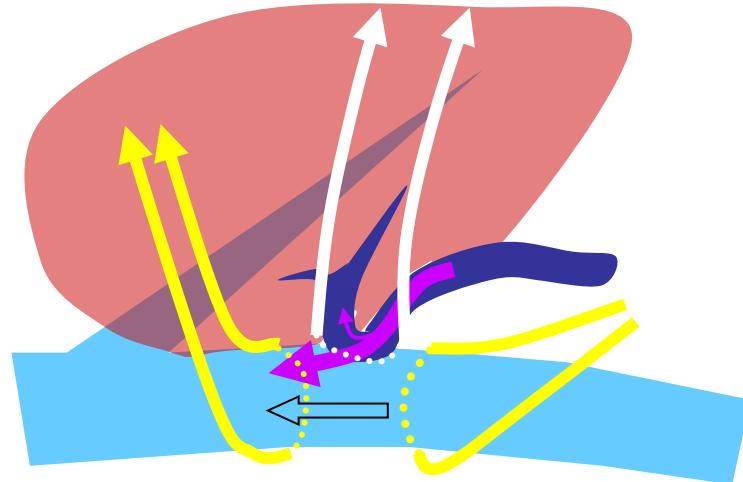
# Monitoring



# Se méfier des faux types!

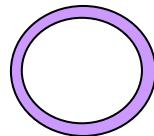
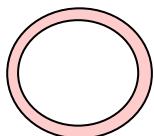


2 - step  
: banding ?

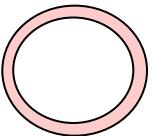


# Comment s'en assurer?

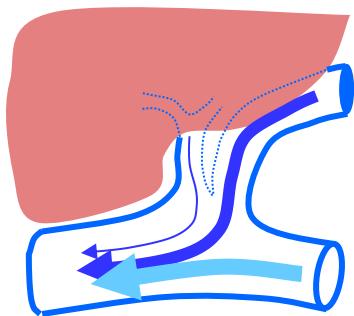
Small  
Bowel  
appearance



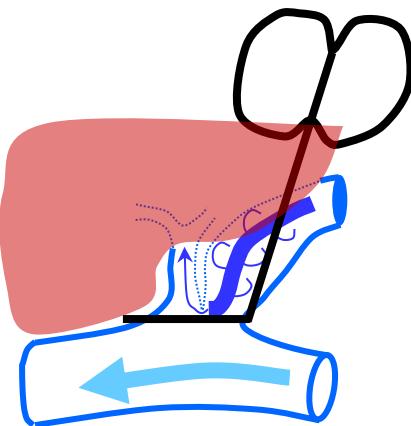
— 15 mn →



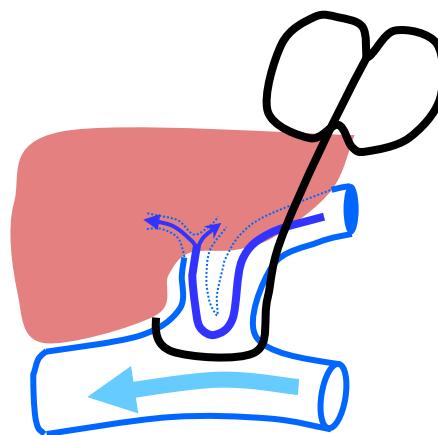
Portal  
blood  
pressure



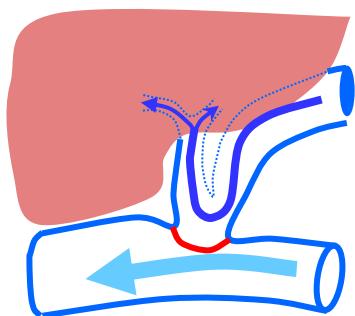
a



b

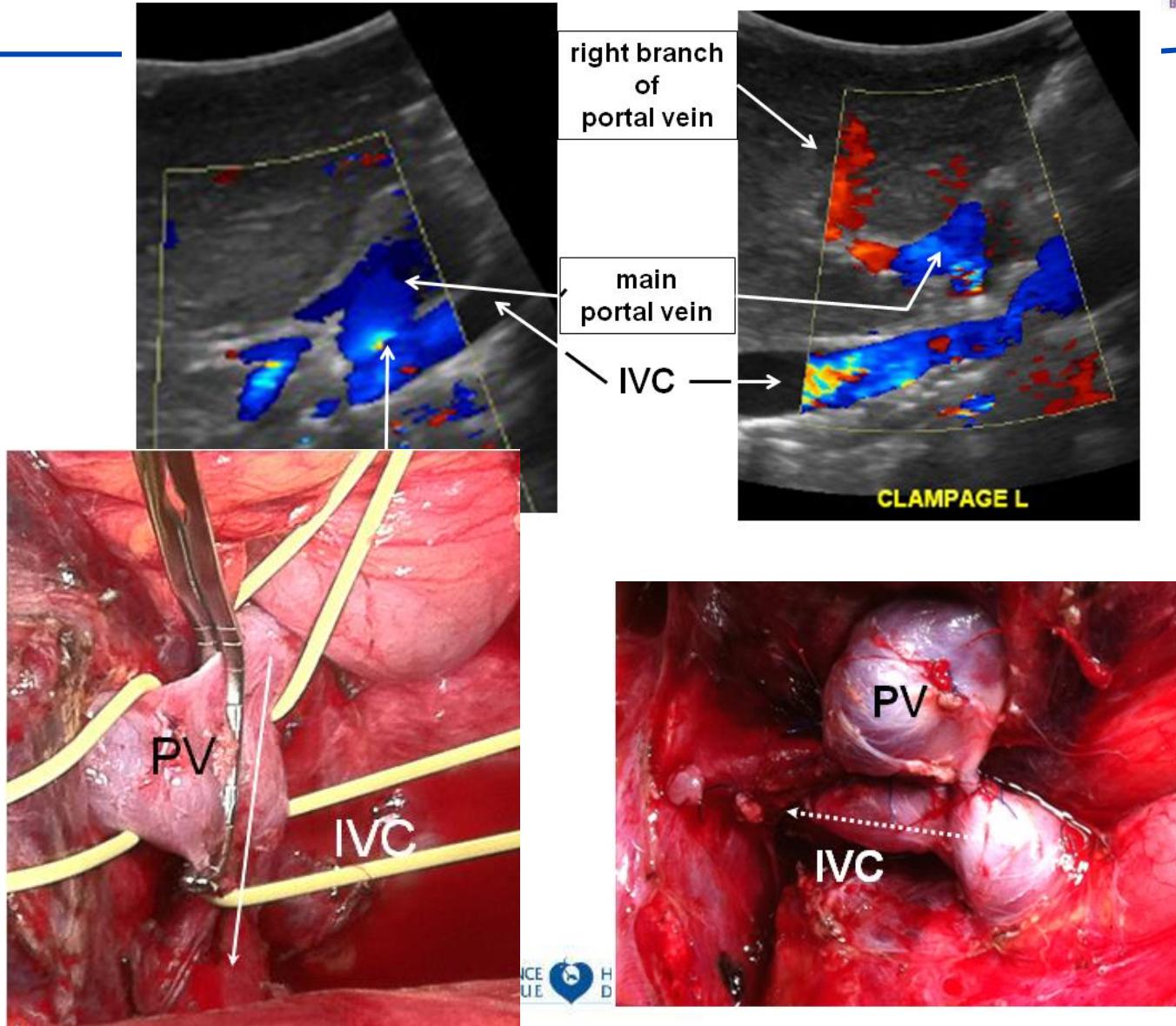


c

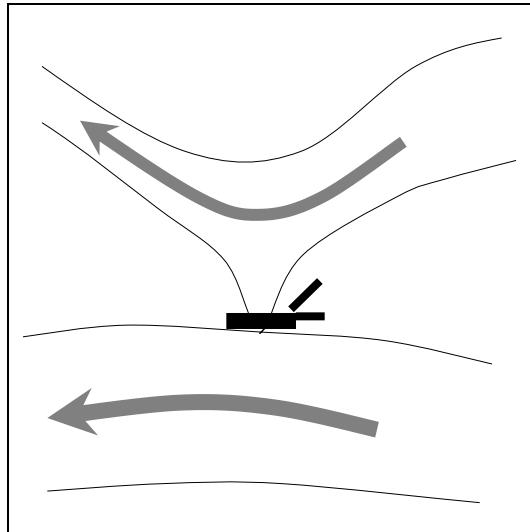


d

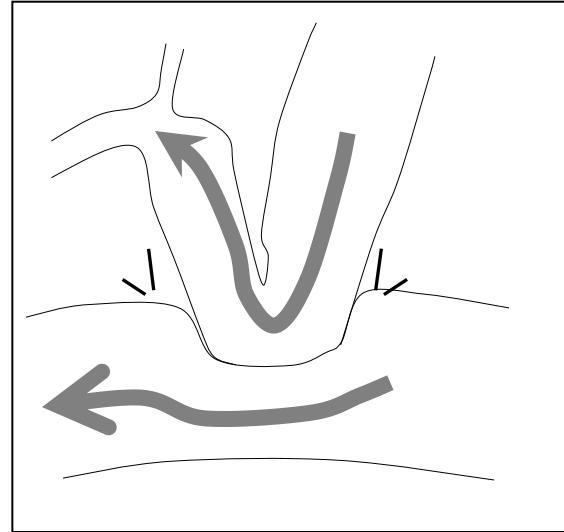
# Monitorage Doppler



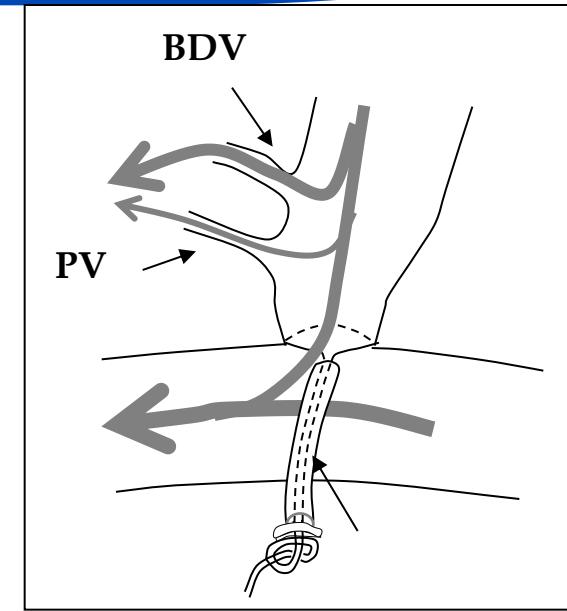
# Différents types de fermeture



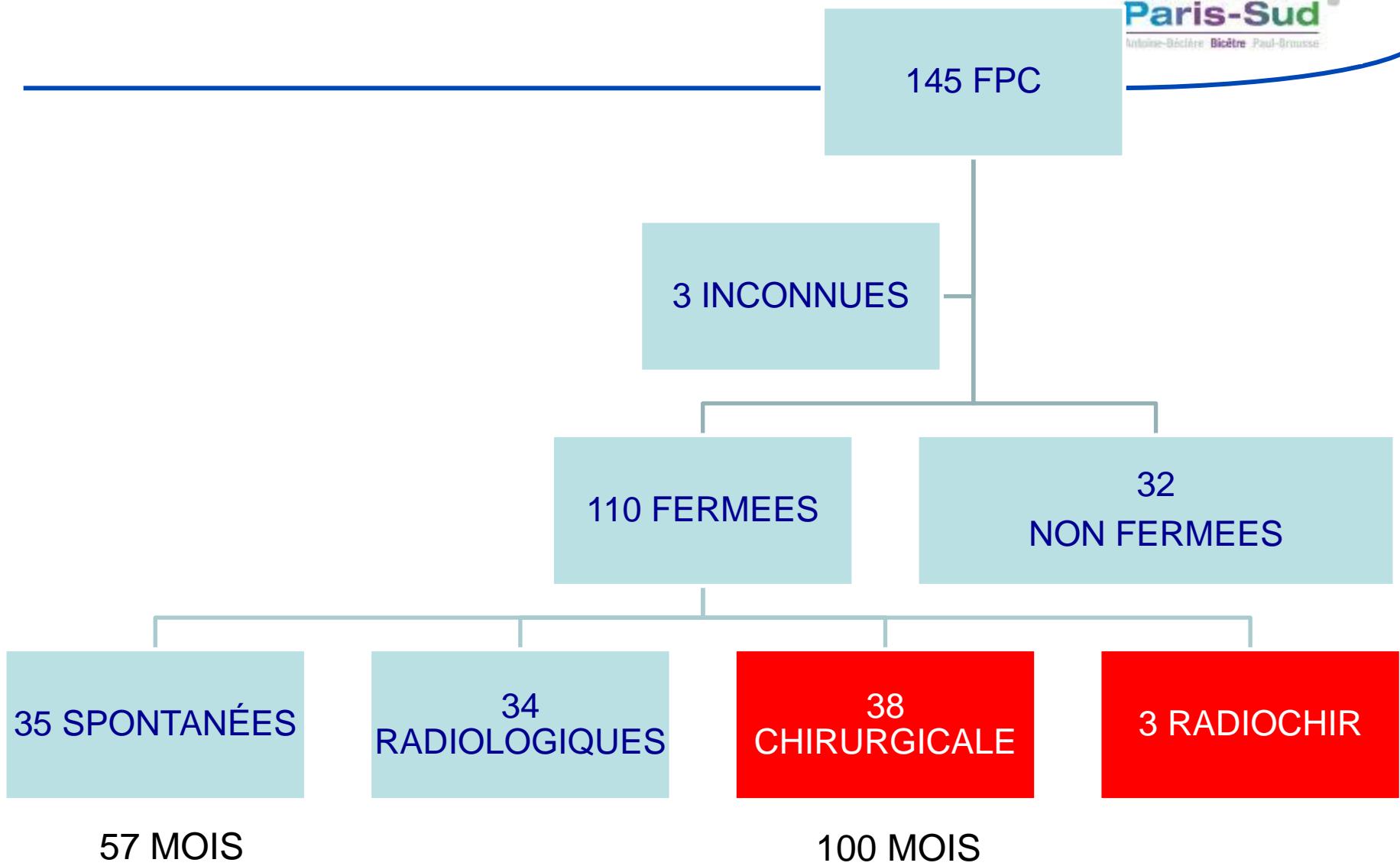
Ligation

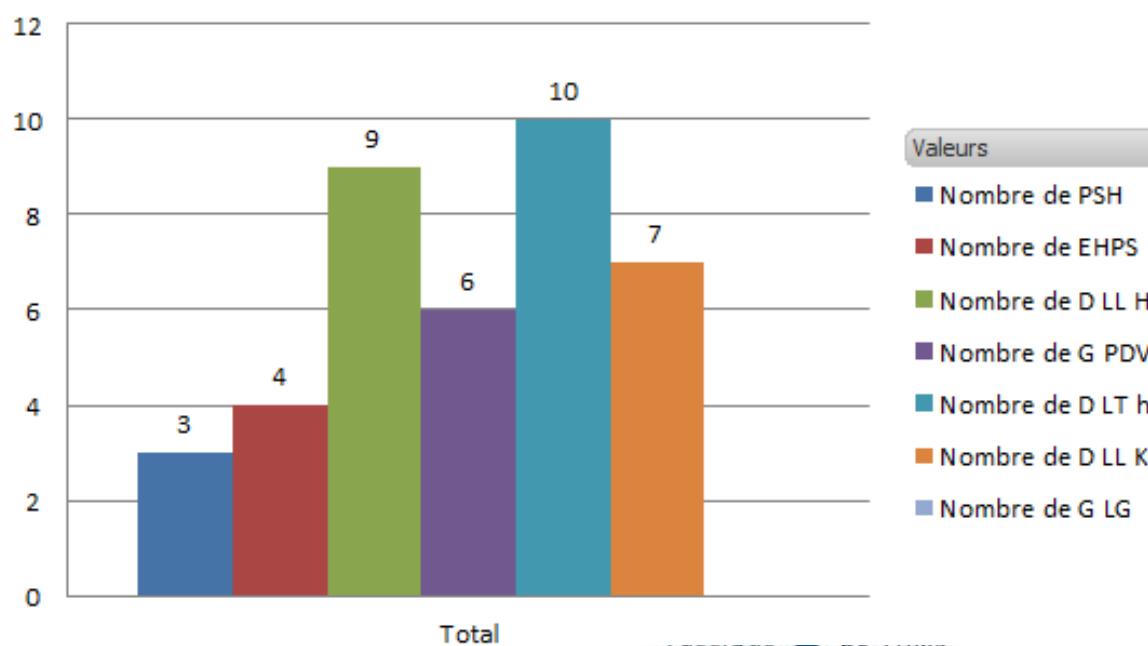
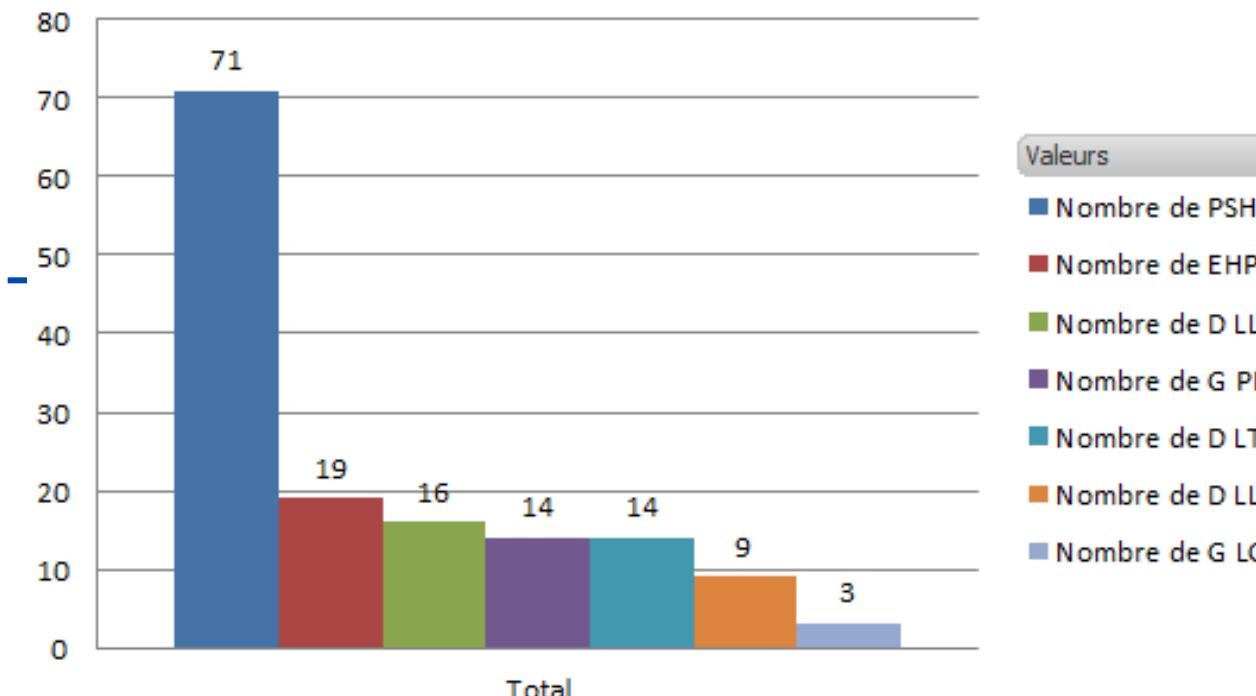


Caval partition

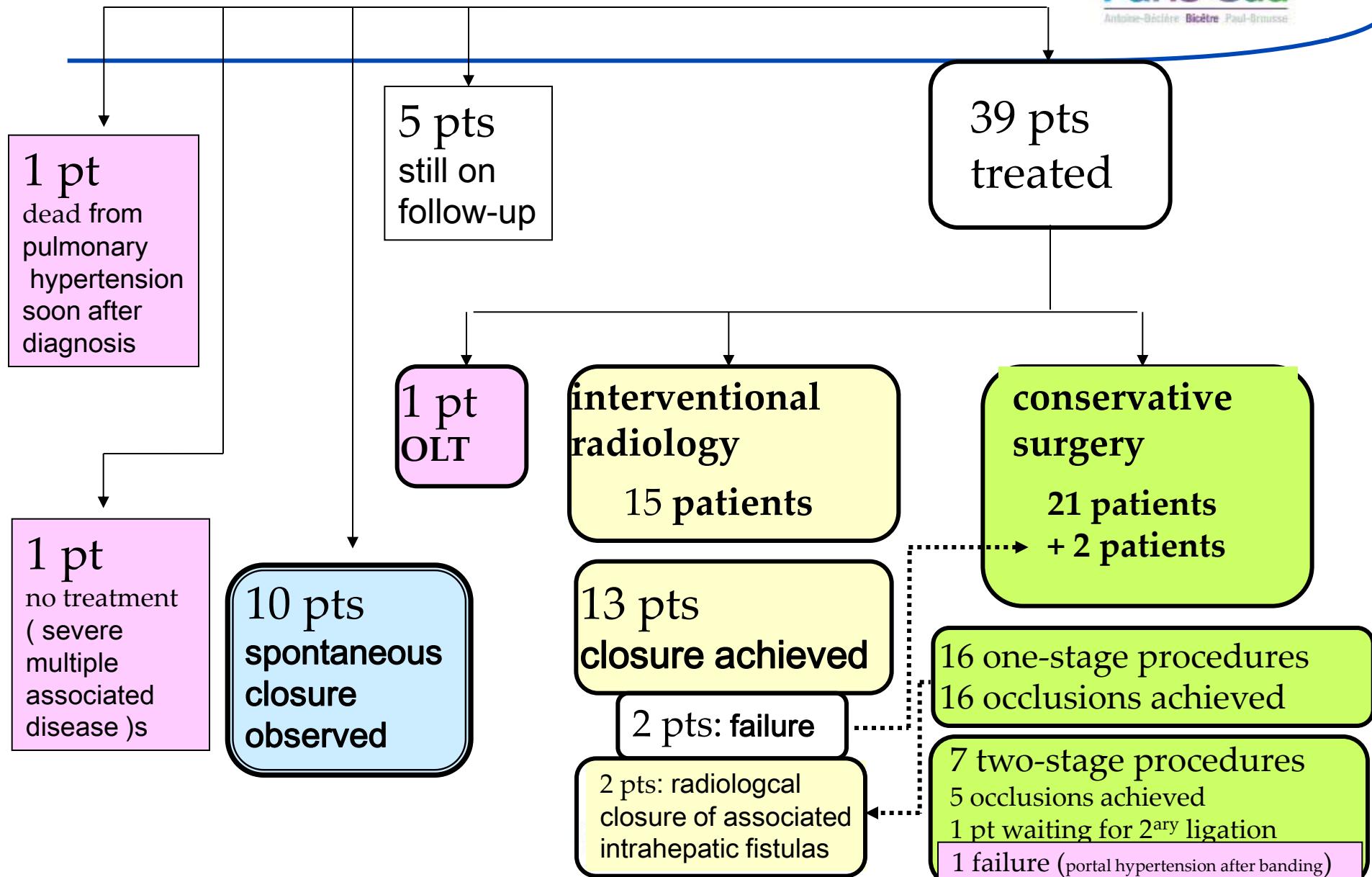


**Banding**  
Fermeture différée





# 56 patients Bicêtre for PC / PHV fistulas



# La suite: USC

---

↘ Pas d'ascite

↘ Echo doppler J0-J1:

- Thrombose
- Ascite

↘ Alimentation rapide

↘ Habituellement:

- Atélectasies
- VNI
- Shunts+++

# Suite en Hospi

---

- ~ Anticoagulants jusqu'à J7-J10
- ~ Post opératoire
  - Acides biliaires
  - Ammoniémie post et à Jeun:+/-
- ~ Consultation précoce J21
- ~ 3, 6 mois
- ~ Bilan post fermeture

# Cas particulier : fermetures en deux temps

- >Type de Fistule: h
- ~ Ou.. Complex: parfois exceptions
  - Intérêt du monitorage
  - Intérêt de la classif (monitorages limites)
- ~ Se mettre à 6 mois un an
- ~ Équipes proposant double temps précoce
  - Ascite
  - Lourdeur?

# Les nodules

## ➤ Bénins

- Laisser++
- Régression
- Mais Adénomes HC  $\beta$ -mutés??

## ➤ Malins: Adultes

- Résection concomitante?
  - Anticoagulants
  - Risques oncologiques et chir
- Transplantation?
  - Risque de récidives CHC

# Nodules dg sur Douleur abdo



Biopsie: adénome bCat activé

11 ans

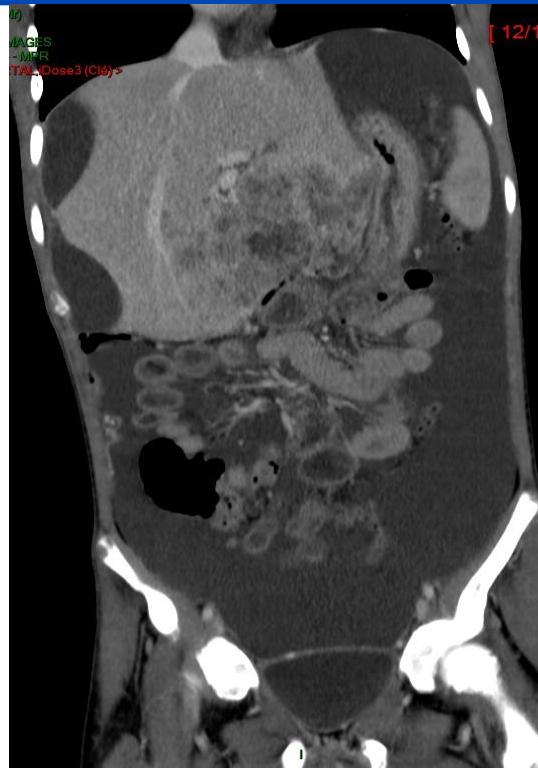
Shunt Droit Termino latéral (Abernethy 1)

Fermeture en deux temps sans complications

2 nodules

- **Plus de Nodules à 1 an**

# 2 ans après CHC métastatique

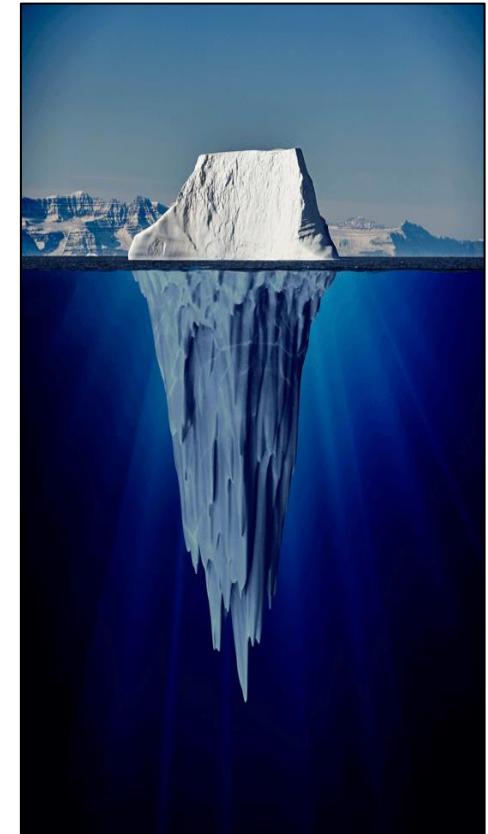


# La transplantation hépatique

- ~ 2 cas/145
- ~ Ne pas croire ni le scanner ni la phlébographie
- ~ Cavernome compensé > immunosuppression et 10% de mortalité à 6 mois?
- ~ Cas réservés aux
  - Tumeurs malignes?
  - Malformation (Fistules complexes intra + extra Hépatique)
  - Complications sévères
- ~ Technique :
  - Branchement VMS
  - Greffon VMS

# Summary

- All ages
- Asymptomatic vs complications
- Multisystem disease
- Suspect!
- Follow up
- Spontaneous closure possible
- Close in symptomatic patients
- More to come!



# International Registry of Congenital Porto-Systemic Shunts

## ESPGHAN Working Group

- Open to all
- PIs:
  - Pr Valérie McLin [valerie.mclin@hcuge.ch](mailto:valerie.mclin@hcuge.ch)
  - Dr Stéphanie Franchi-Abella [stephanie.franchi@aphp.fr](mailto:stephanie.franchi@aphp.fr)
  - Pr Emmanuel Gonzales [emmanuel.gonzales@aphp.fr](mailto:emmanuel.gonzales@aphp.fr)
  - Pr Dominique Debray [dominique.debray@aphp.fr](mailto:dominique.debray@aphp.fr)
- Research coordinator: Dr Simona Korff [simona.korff@hcuge.ch](mailto:simona.korff@hcuge.ch)

## Multicenter, international collaboration

- Retrospective and prospective
- Ethics approval
- Database - server at
- Images - server at