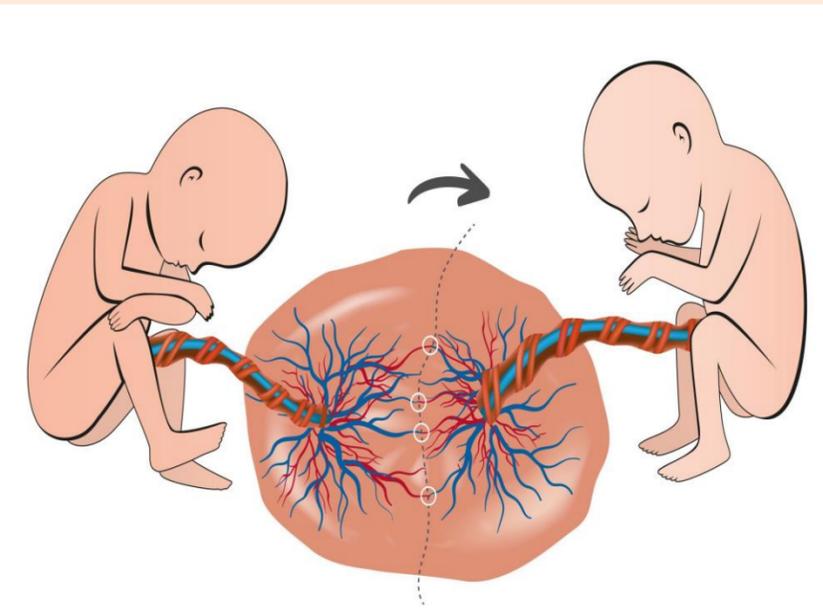


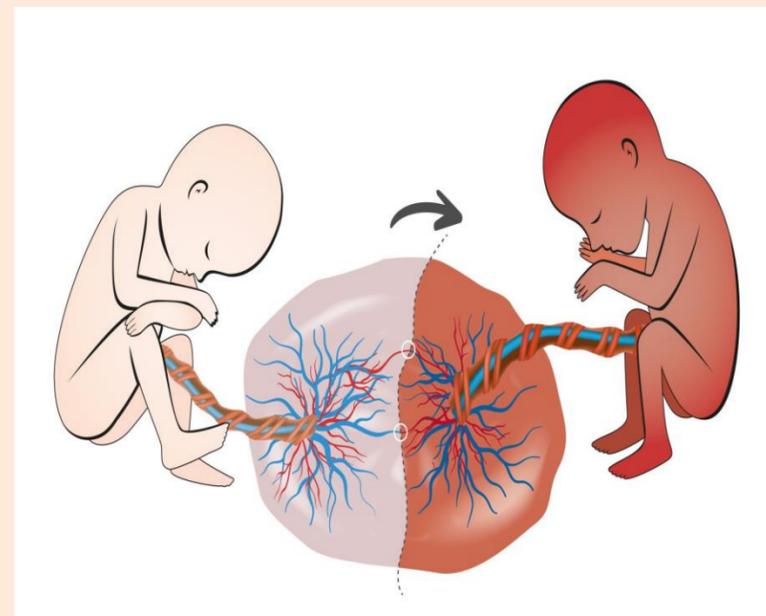
PEC thérapeutique des complications monochoriales



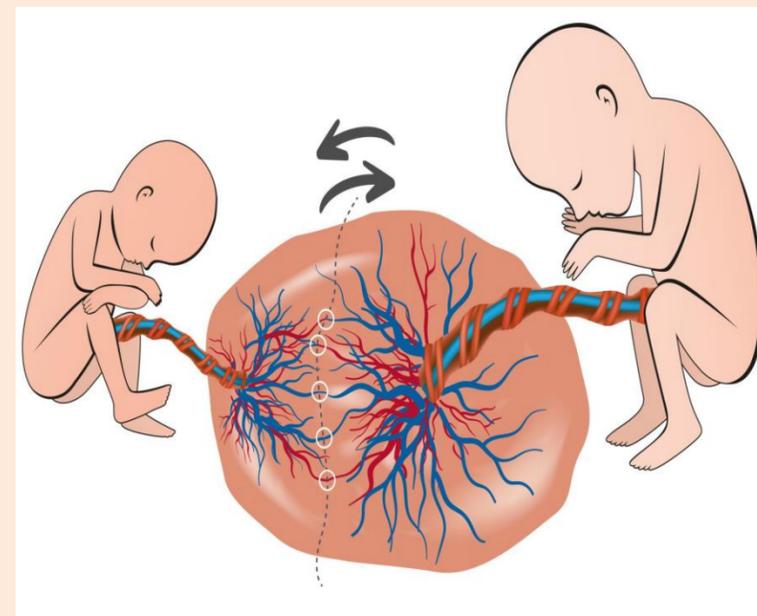
Journée du CPDPN 13.12.2024
Marie ZILLIOX - JAMET



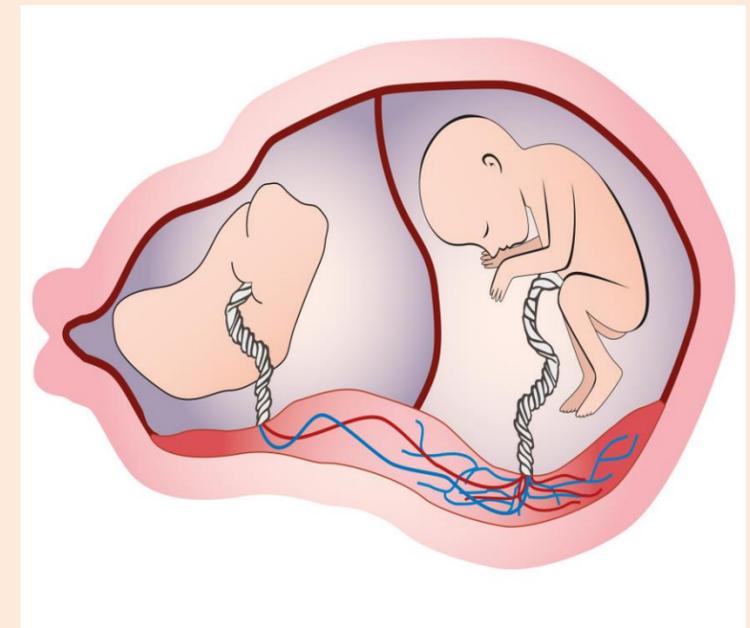
STT



TAPS



RCIU sélectif

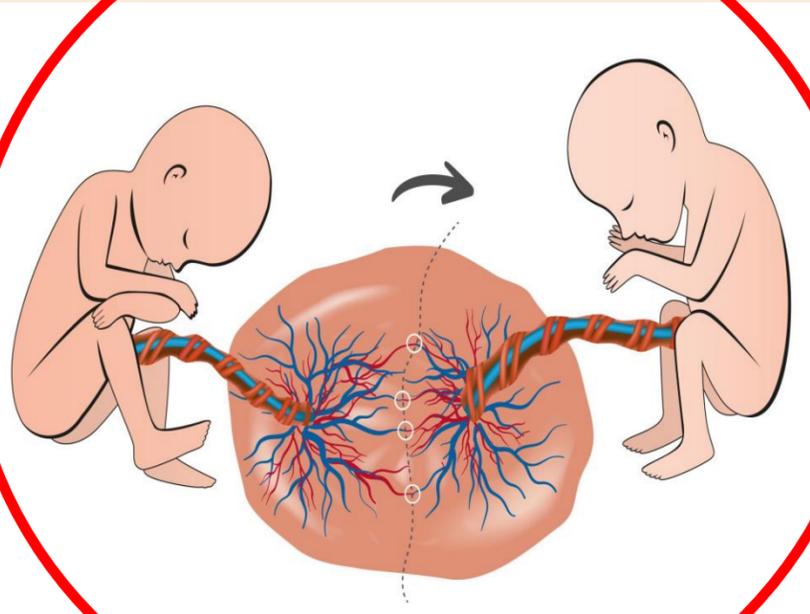


TRAP

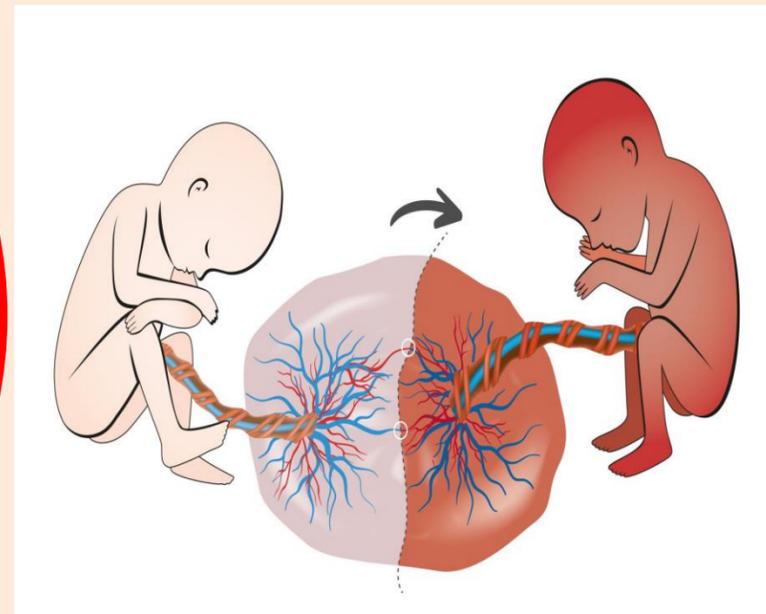
PEC thérapeutique des complications monochoriales



Journée du CPDPN 13.12.2024
Marie ZILLIOX - JAMET



STT



TAPS

Recommandations de la SMFM

Octobre 2024 !

UNNUMBERED TABLE
Summary of recommendations

| Number | Recommendation | GRADE |
|--------|--|---------------|
| 1 | We recommend routine first-trimester sonographic determination of chorionicity and amnionity. | 1B |
| 2 | We recommend that ultrasound surveillance for TTTS begin at 16 weeks of gestation for all MCDA twin pregnancies and continue at least every 2 weeks until delivery, with more frequent monitoring indicated with clinical concern. | 1C |
| 3 | We recommend that routine sonographic surveillance for TTTS minimally include assessment of amniotic fluid volumes on both sides of the intertwin membrane and evaluation for the presence or absence of urine-filled fetal bladders, and ideally incorporate Doppler study of the umbilical arteries. | 1C |
| 4 | We recommend fetoscopic laser surgery as the standard treatment for stage II through stage IV TTTS presenting between 16 and 26 weeks of gestation. | 1A |
| 5 | We recommend expectant management with at least weekly fetal surveillance for asymptomatic patients continuing pregnancies complicated by stage I TTTS and consideration for fetoscopic laser surgery for stage I TTTS presentations between 16 and 26 weeks of gestation complicated by additional factors such as maternal polyhydramnios-associated symptomatology. | 1B |
| 6 | We recommend an individualized approach to laser surgery for early- and late-presenting TTTS. | 1C |
| 7 | We recommend that all patients with TTTS qualifying for laser therapy be referred to a fetal intervention center for further evaluation, consultation, and care. | Best Practice |
| 8 | After laser therapy, we suggest weekly surveillance for 6 weeks followed by resumption of every-other-week surveillance thereafter, unless concern exists for post-laser TTTS, post-laser TAPS, or FGR. | 2C |
| 9 | Following the resolution of TTTS after fetoscopic laser surgery and without other indications for earlier delivery, we recommend delivery of dual-surviving MCDA twins at 34 to 36 weeks of gestation. | 1C |
| 10 | In TTTS pregnancies complicated by posttreatment single fetal demise, we recommend full-term delivery (39 weeks) of the surviving co-twin to avoid complications of prematurity unless indications for earlier delivery exist. | 1C |
| 11 | We recommend that fetoscopic laser surgery not influence the mode of delivery. | Best Practice |
| 12 | We recommend that prenatal diagnosis of TAPS minimally require either Doppler MCA-PSV values >1.5 MoM and <1.0 MoM in donor and recipient twins, respectively, or an intertwin Δ MCA-PSV >0.5 MoM. | 1C |
| 13 | We recommend that providers consider incorporating Doppler MCA-PSV determinations into all MC twin ultrasound surveillance beginning at 16 weeks of gestation. | 1C |
| 14 | Consultation with a specialized fetal care center is recommended when TAPS progresses to a more advanced disease stage (\geq II) before 32 weeks of gestation or when concern arises for coexisting complications such as TTTS. | Best Practice |

Quand? Qui?

TERME

Seuils = 16/17 SA – 26 SA

STADE

Stade 1 si symptomatologie / complications
/ évolution

Stade 2 +++

Traitement

Comment?

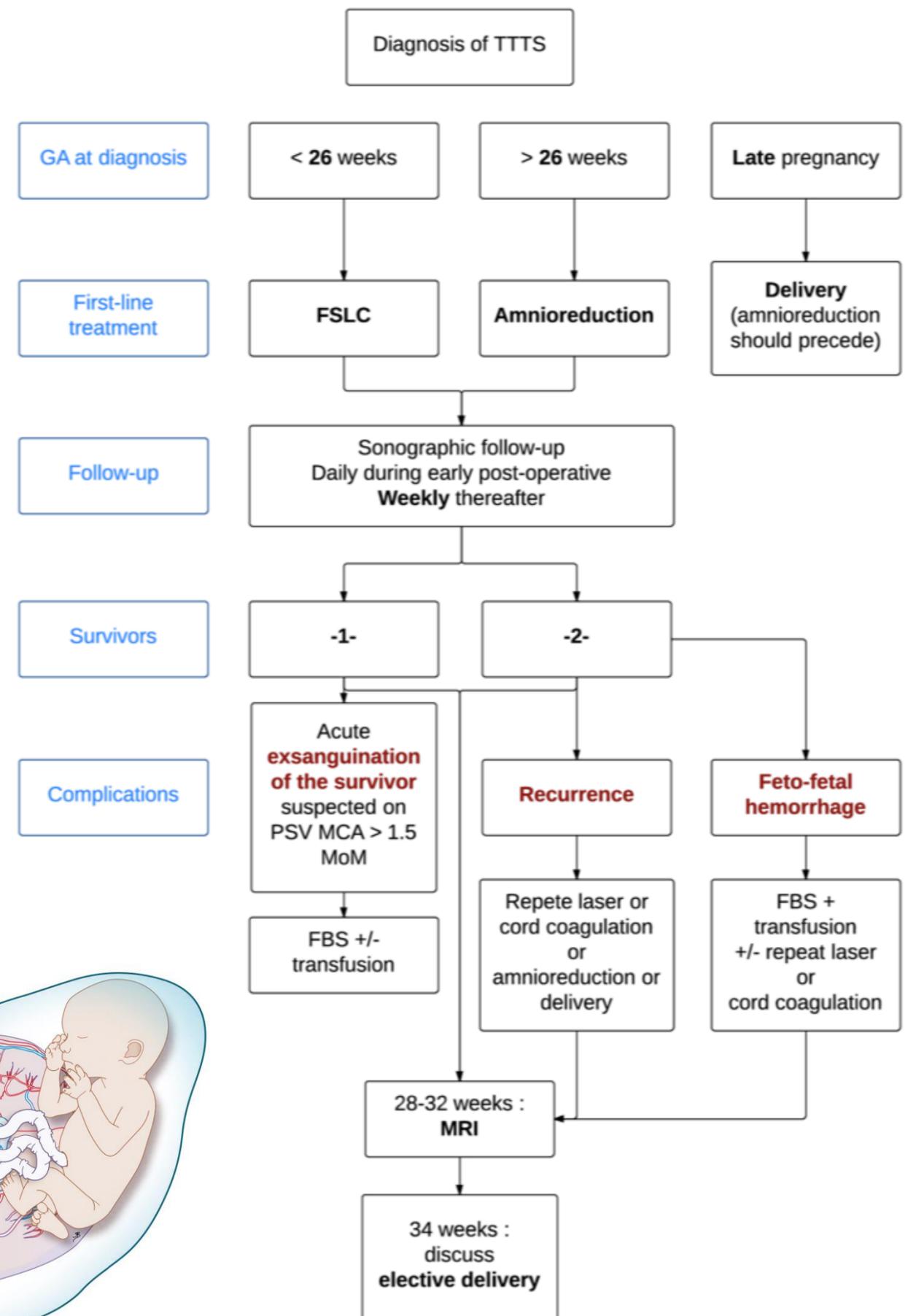
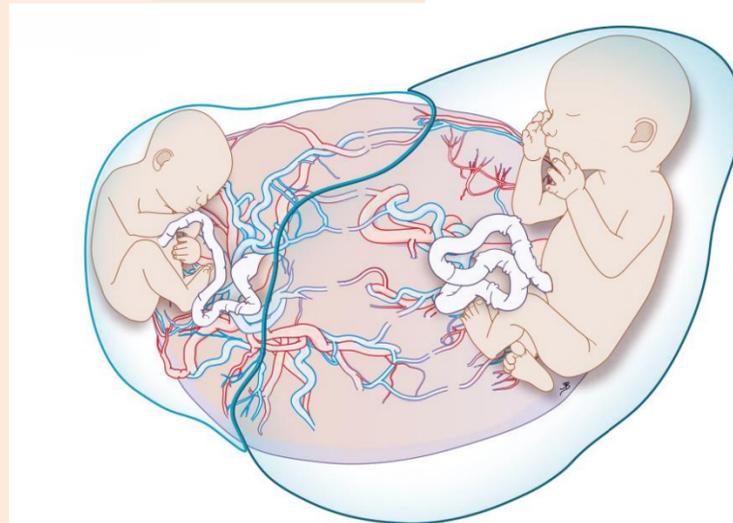
- 1. Foetoscopie laser**
= gold standard et seul traitement étiologique
- 2. Occlusion de cordon ombilical : à défaut**
 - échec de foetoscopie laser
 - RCIU associé
- 3. Amniodrainage**
Traitement symptomatique
- 4. Accouchement**
Traitement symptomatique

Quand et qui traiter ?

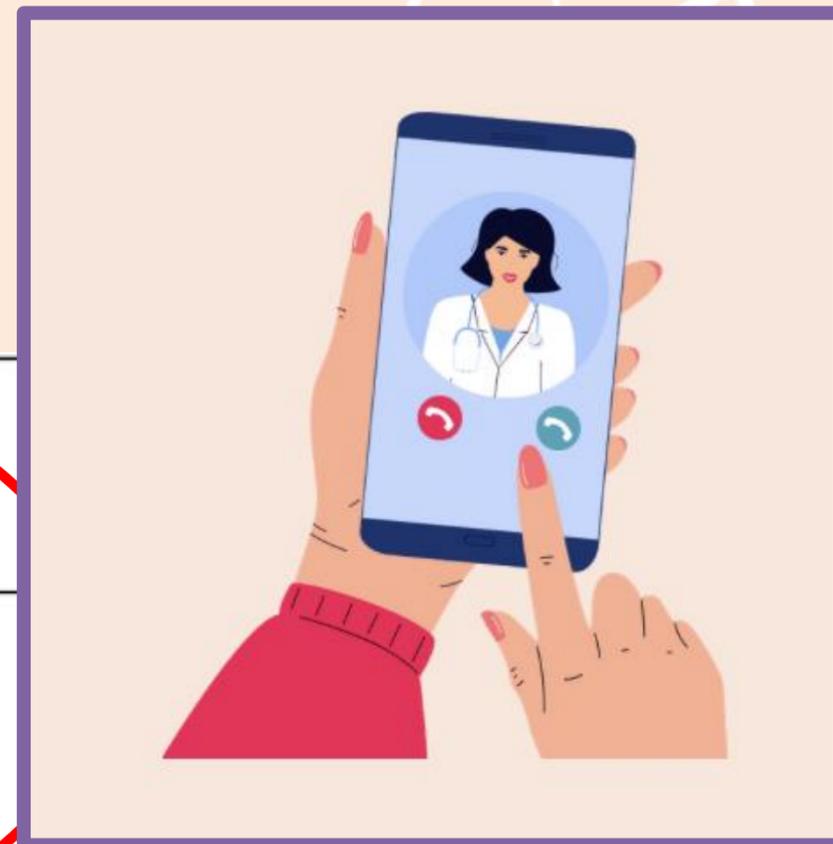
- Dès suspicion diagnostique : adresser en centre de reference
- **Stade 1 ?**
 - **Suivi hebdomadaire**
 - 60% évolutifs = intervention recommandée
 - Parmi 40% regression : >90% de survie, et taux de complications neurologiques faible

• A partir du stade 2

- B/R en faveur d'une **intervention +++**
- Entre 16 et 26 SA



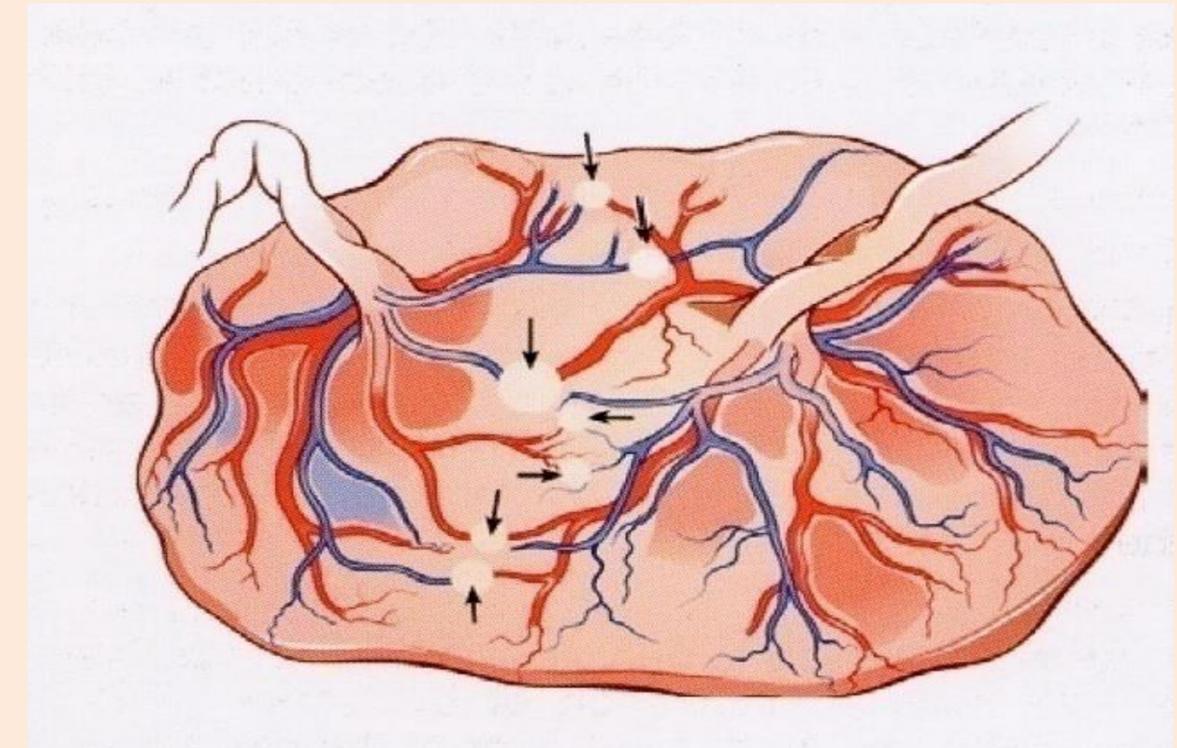
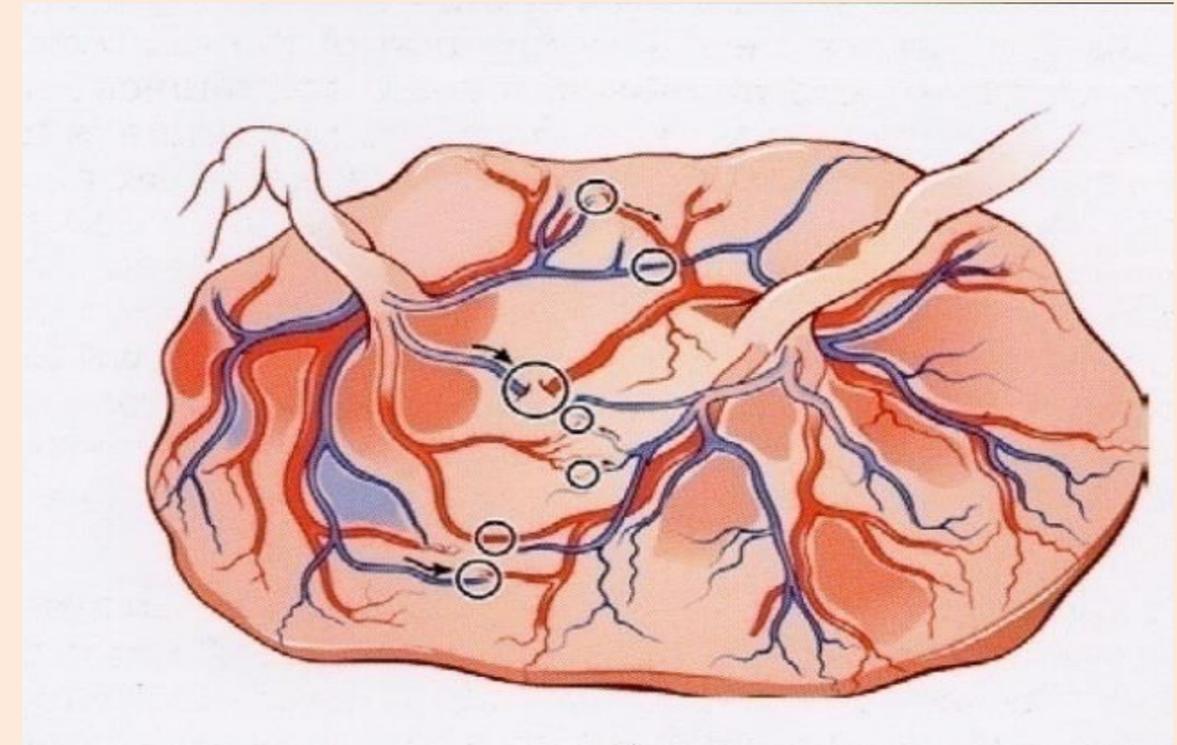
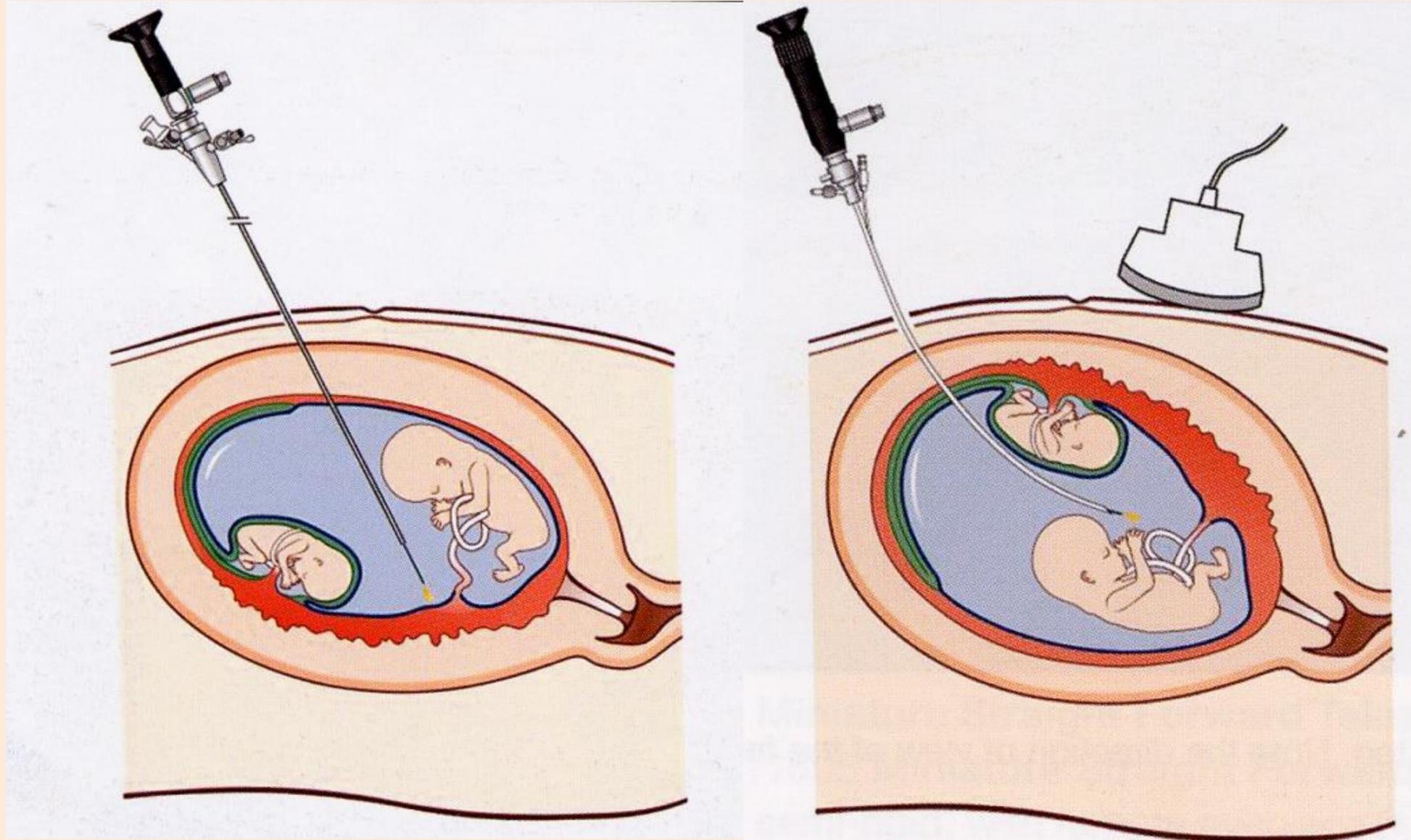
Quand et qui traiter ?



| | | |
|---|--|---------------|
| 4 | We recommend fetoscopic laser surgery as the standard treatment for stage II through stage IV TTTS presenting between 16 and 26 weeks of gestation. | |
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| 6 | We recommend an individualized approach to laser surgery for early- and late-presenting TTTS. | 1C |
| 7 | We recommend that all patients with TTTS qualifying for laser therapy be referred to a fetal intervention center for further evaluation, consultation, and care. | Best Practice |

La foetoscopie avec coagulation laser

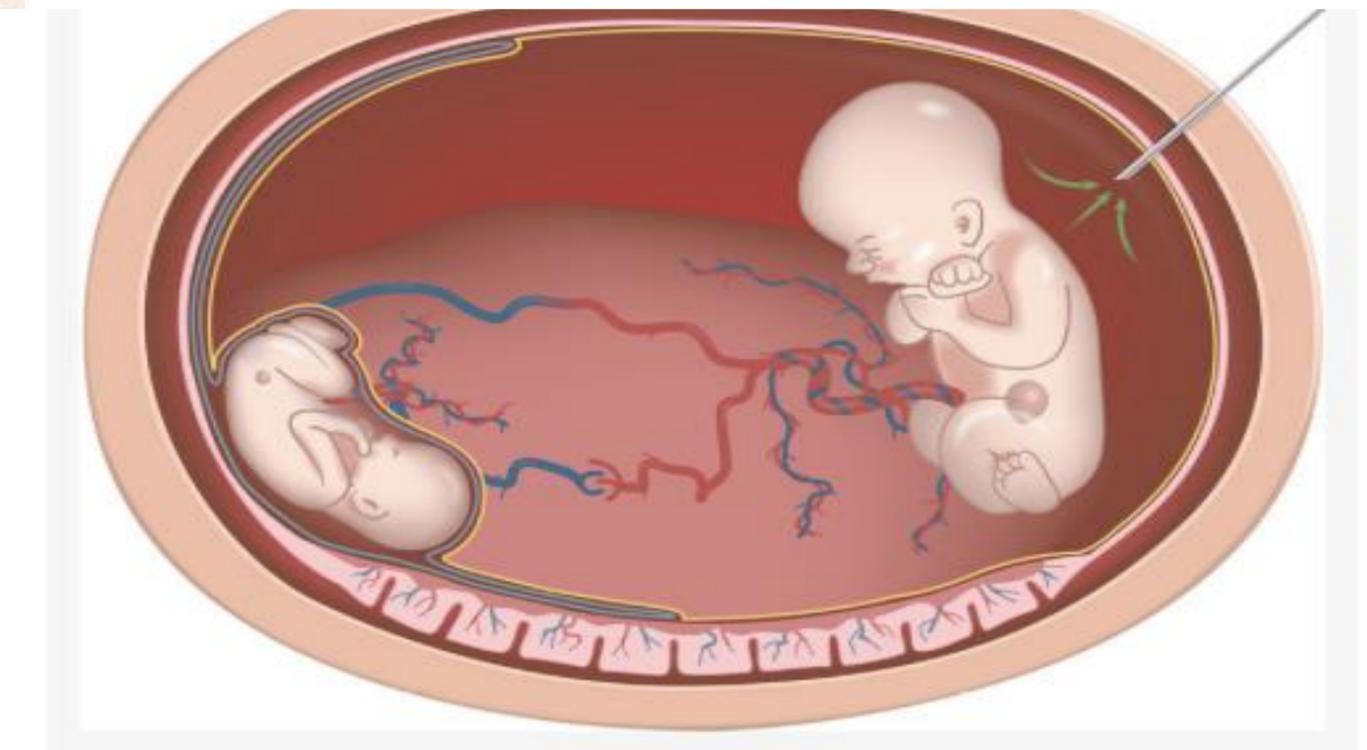
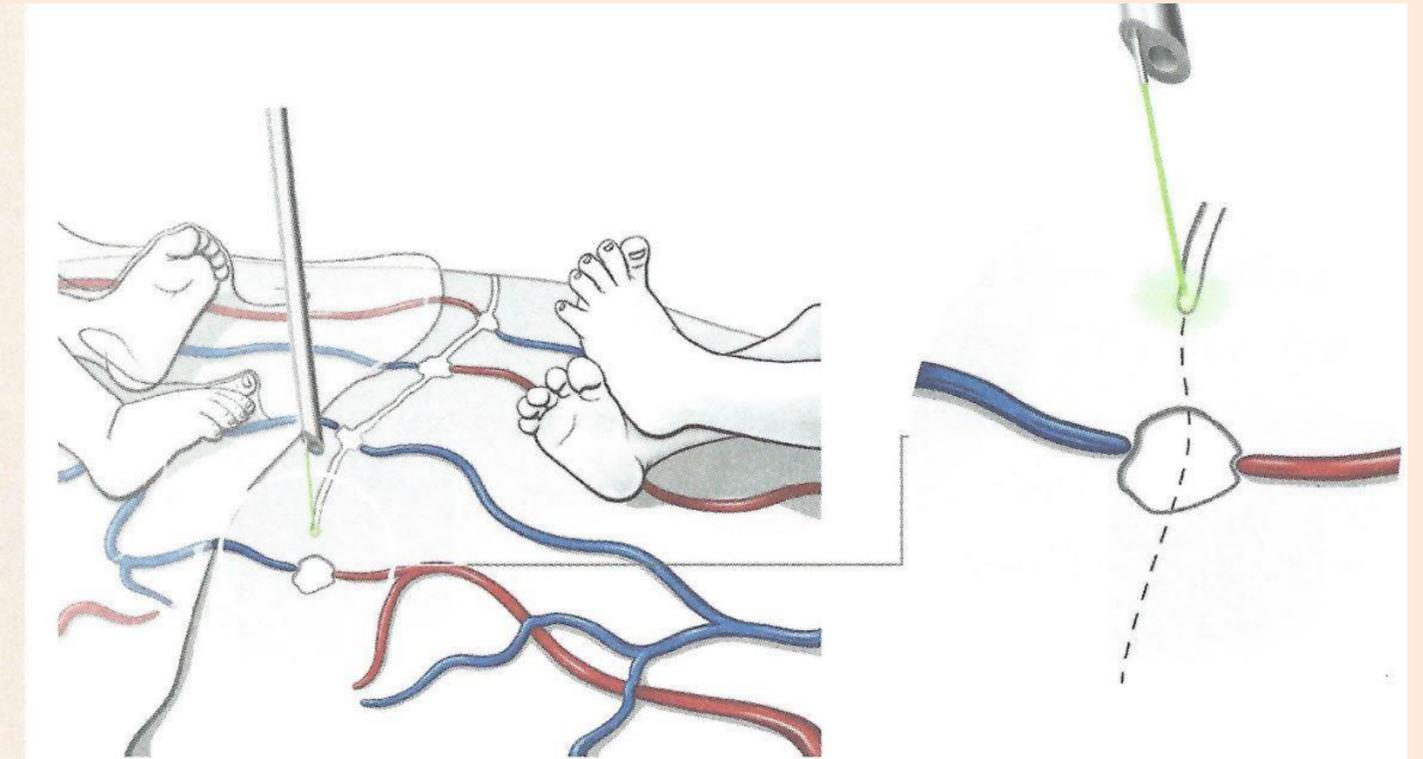
Abord percutané



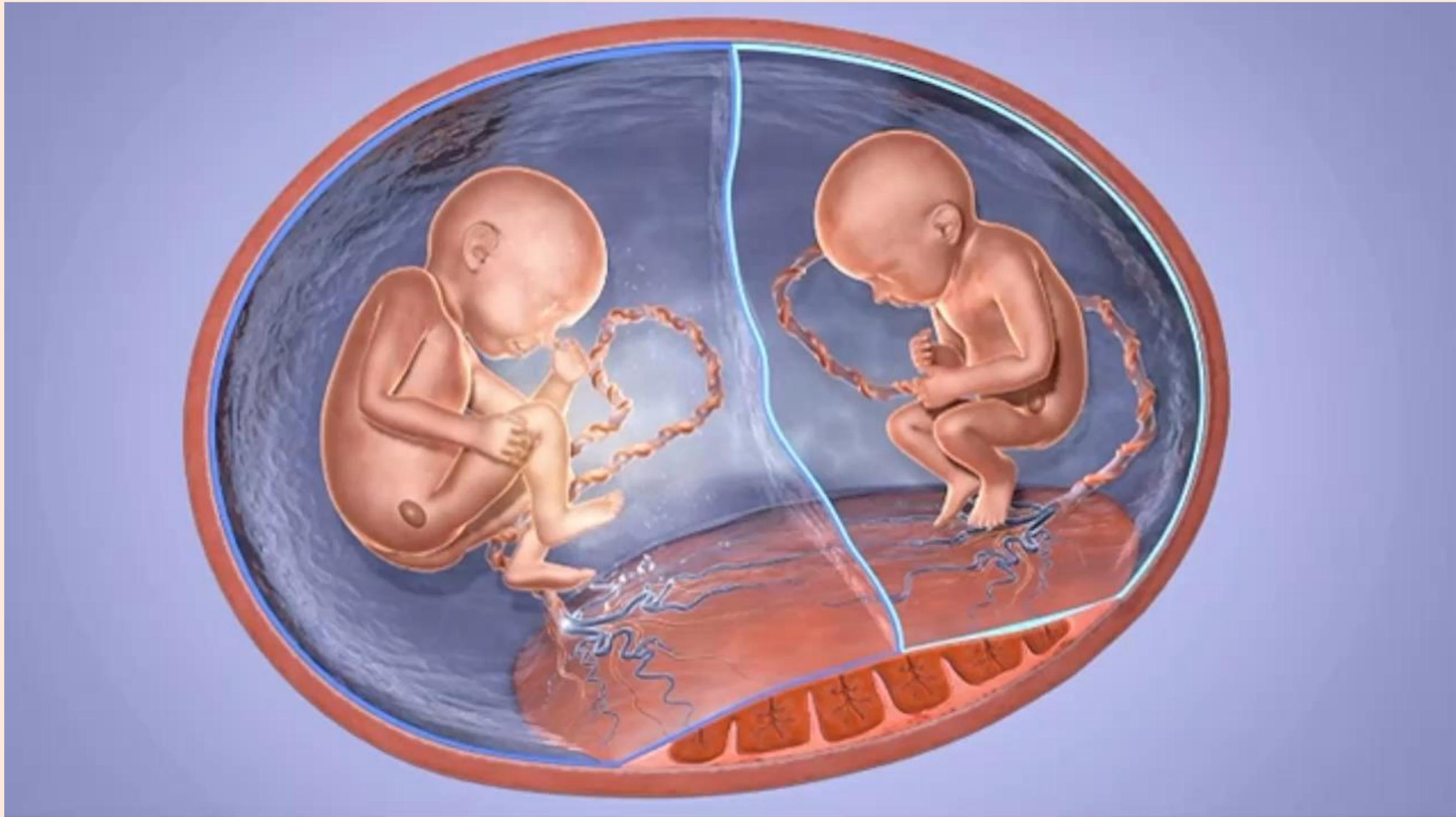
Sous sédation maternelle IV et AL

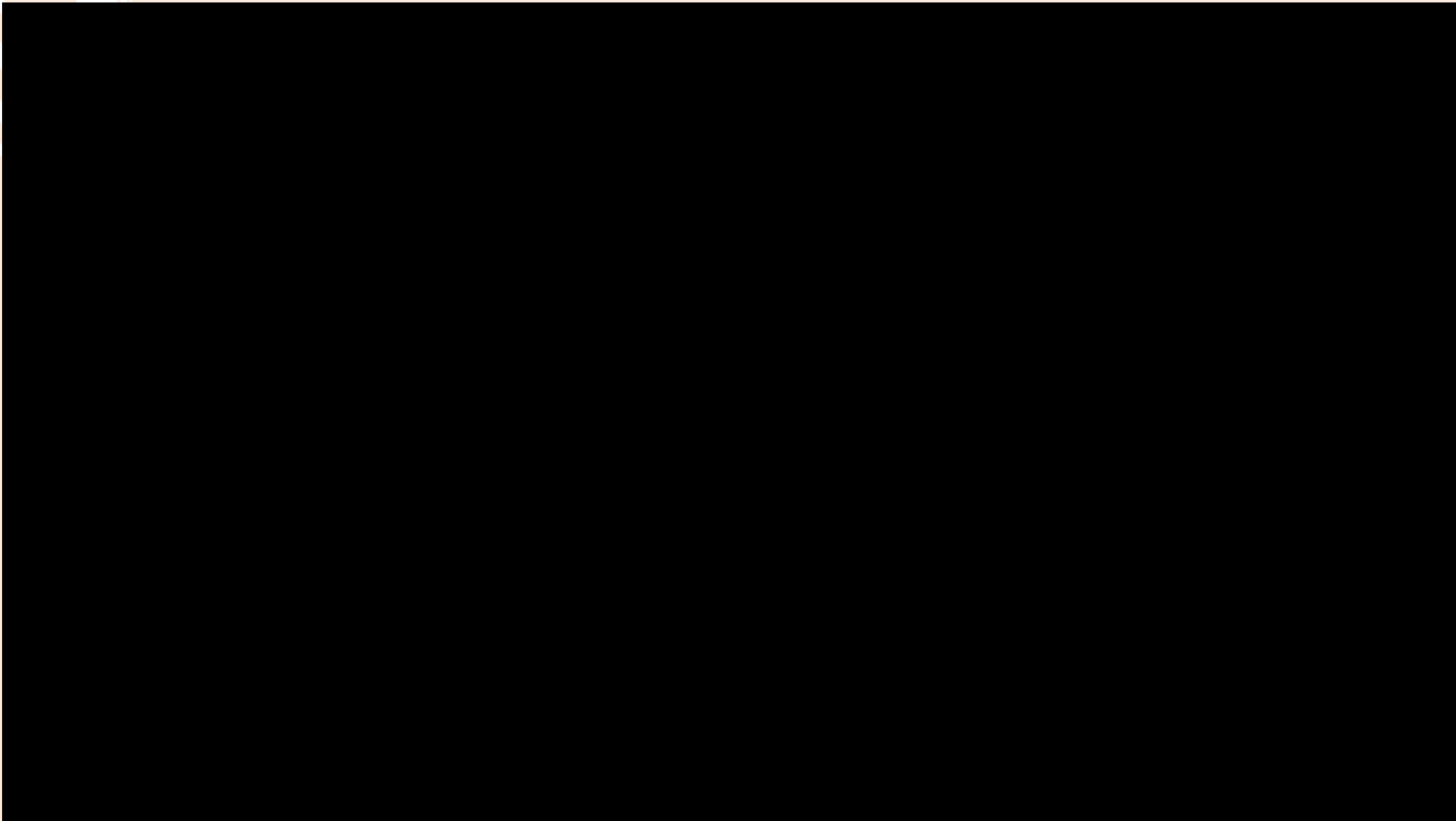
Traitement par foetoscopie laser

- Traitement de référence = **gold standard**
- **Technique de Solomon = bichorionisation placentaire**
- Alternatives thérapeutiques :
 - **Amniodrainage = ttt** symptomatique
 - **ISG** par occlusion du CO
 - **Accouchement**

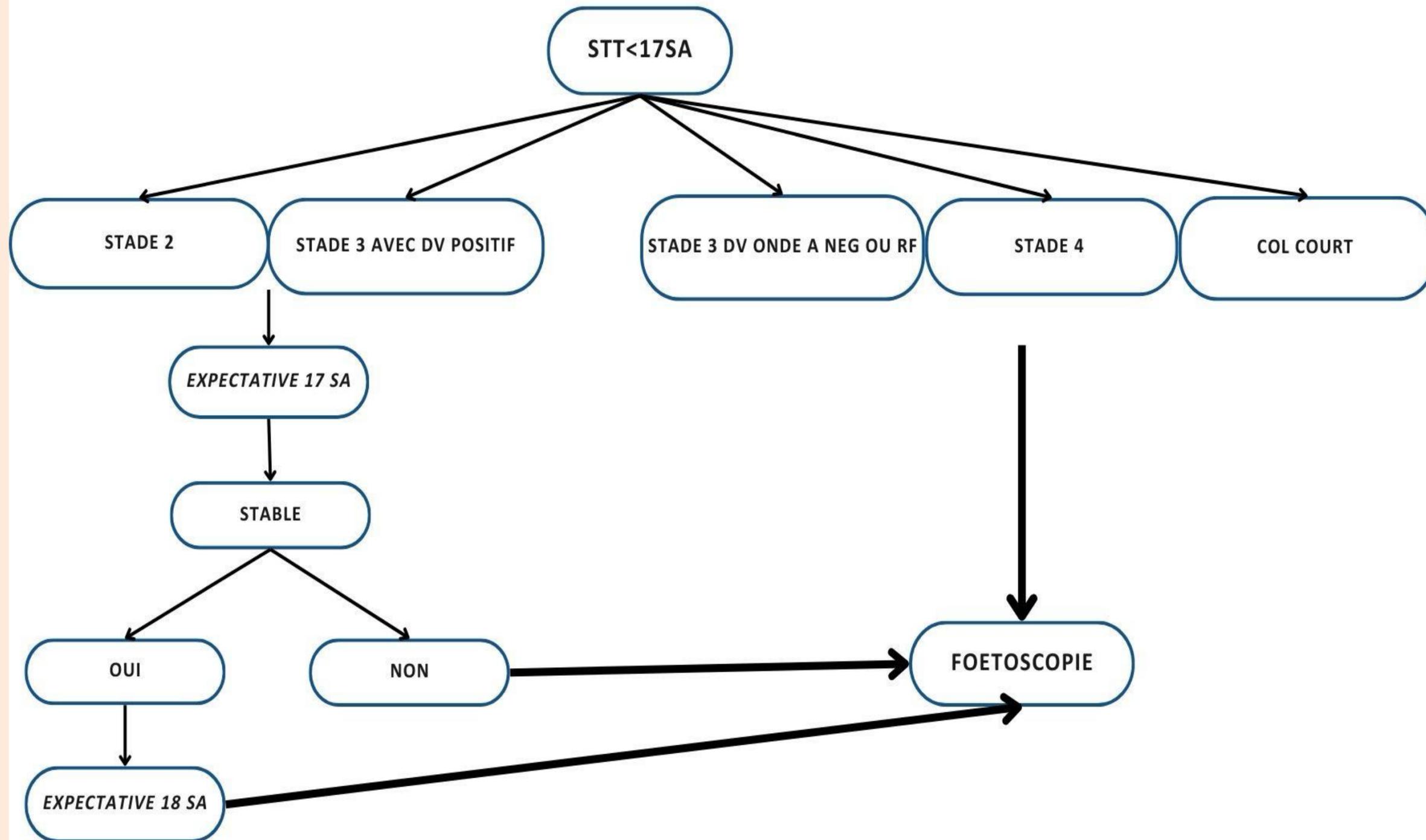


Le Syndrome Transfuseur Transfusé (TOPS)

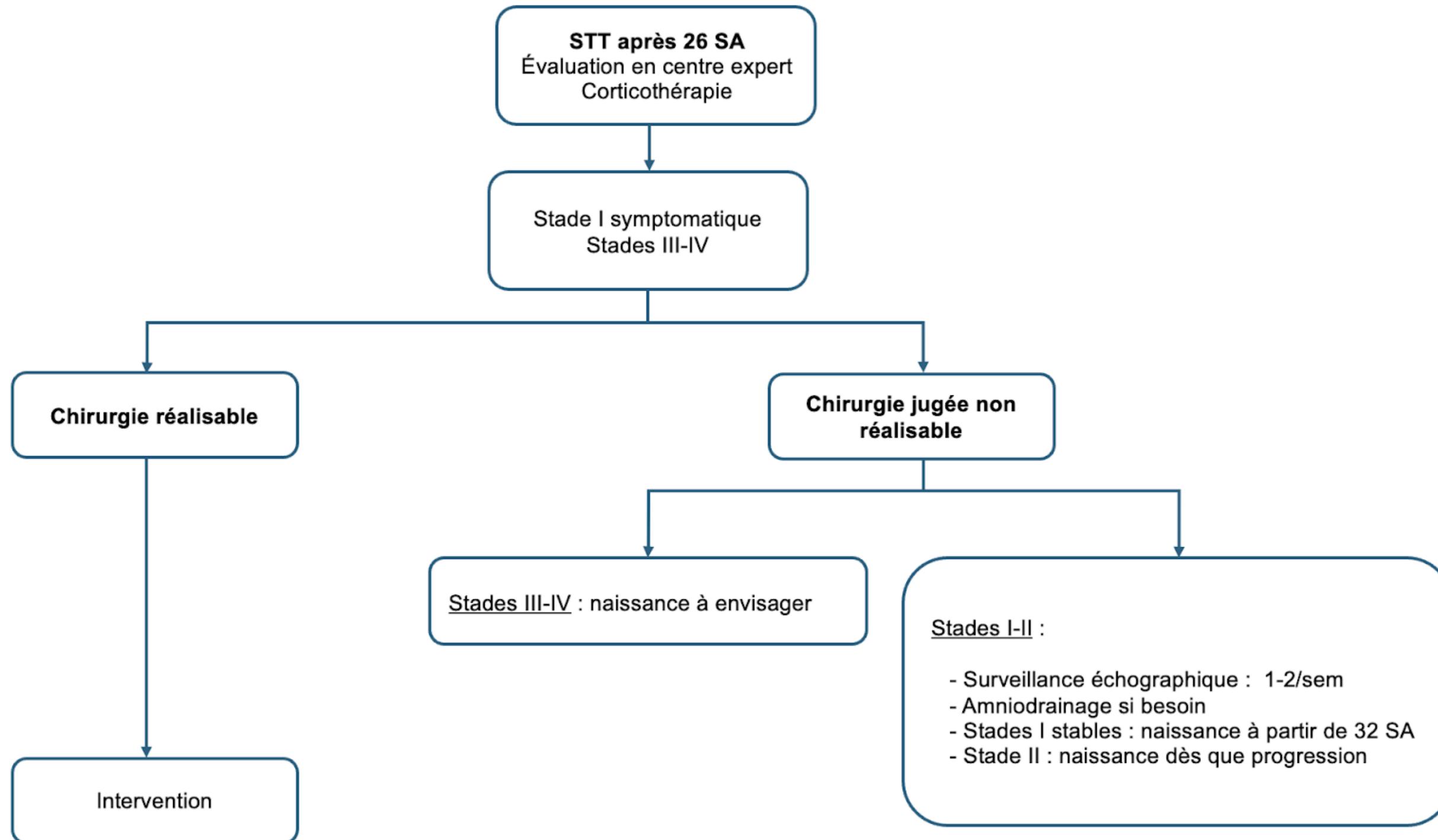




Ages gestationnels précoces



Ages gestationnels tardifs



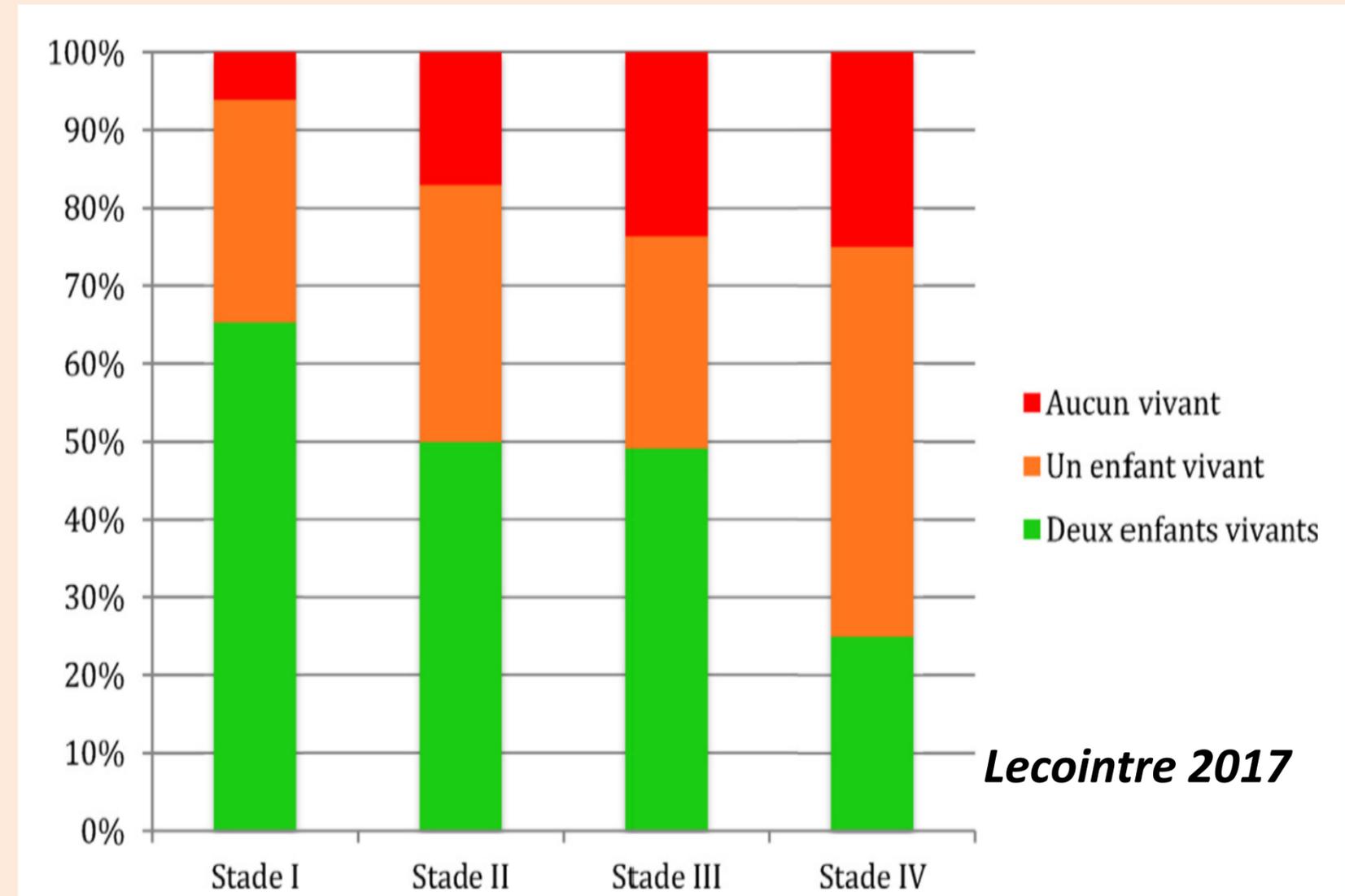
Résultats après foetoscopie



| | Overall, N = 298 ¹ |
|----------------|-------------------------------|
| 0 | 23 (7.8%) |
| 1 | 60 (20%) |
| 2 | 210 (72%) |
| <u>Unknown</u> | 5 |



SMFM 2024



A Strasbourg, en 2017...

¹Statistics presented: n (%)

Facteurs pronostiques



| | | Au moins un enfant vivant | Aucun enfant vivant | p |
|--|-----|---------------------------|---------------------|-------|
| Caractéristiques maternelles et obstétricales | | | | |
| Age de la patiente (années) | | 30.2 ± 5.4 | 28.7 ± 6.2 | 0.22 |
| Parité | 0 | 60 (78.9%) | 16 (21.1%) | 0.18 |
| | ≥ 1 | 107 (86.3%) | 17 (13.7%) | |
| Stade Quintero | I | 46 (93.9%) | 3 (6.1%) | 0.01 |
| | II | 73 (83.0%) | 15 (17.0%) | |
| | III | 42 (76.4%) | 13 (23.6%) | |
| | IV | 6 (75.0%) | 2 (25.0%) | |
| Longueur cervicale (mm) | | 40.5 ± 10.4 | 34.6 ± 14.1 | 0.006 |
| Placenta antérieur | Oui | 62 (74.7%) | 21 (25.3%) | 0.005 |
| | Non | 105 (89.7%) | 12 (10.3%) | |

Facteurs pronostiques



| Caractéristiques chirurgicales | | Au moins un enfant vivant | Aucun enfant vivant | p |
|--|-----------|---------------------------|---------------------|------|
| Age gestationnel au moment du laser (semaines) | | 20.3 ± 3.0 | 19.3 ± 2.9 | 0.07 |
| Endoscope utilisé | 1.3 mm | 25 (83.3%) | 5 (16.7%) | 0.91 |
| | 2 mm | 142 (83.5%) | 28 (16.5%) | |
| Nombre de vaisseaux coagulés | | 9.7 ± 4.5 | 10.7 ± 4.6 | 0.26 |
| Technique | Sélective | 116 (82.9%) | 24 (17.1%) | 0.50 |
| | Solomon | 51 (85.0%) | 9 (15.0%) | |
| Amniodrainage (mL) | | 1728 ± 1128 | 1687 ± 1035 | 0.84 |
| Durée du laser (min) | | 27.3 ± 10.2 | 27.9 ± 12.1 | 0.78 |

Complications post opératoires

- **RPM (10-15%... 25 % ??)**
 - Fausse couche tardive
 - Ou accouchement **prématuré**

Age médian à l'accouchement : **32 SA**

- **Echec**
- **Récidive** de STT
- **TAPS**
- Brides amniotiques
- HRP / **décollement placentaire**
- Septostomie
- **MFIU**

PREMATURITE:

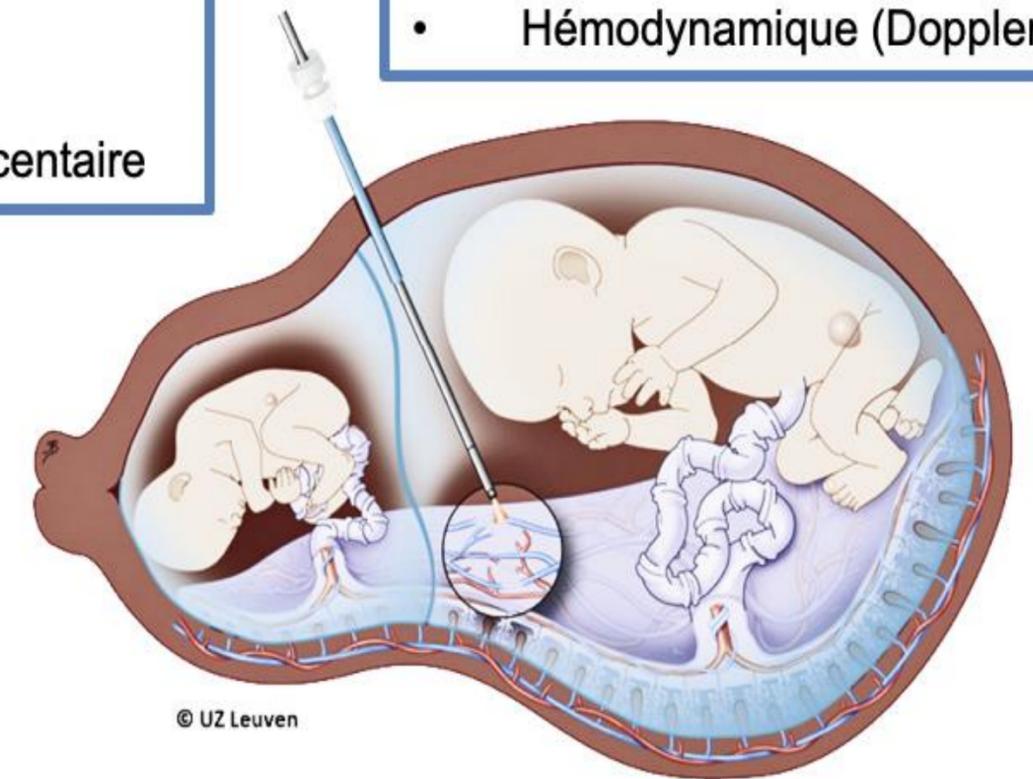
- Col
- RPM

CHIRURGIE:

- Exhaustivité
- Sélectivité
- Technique
- Localisation placentaire

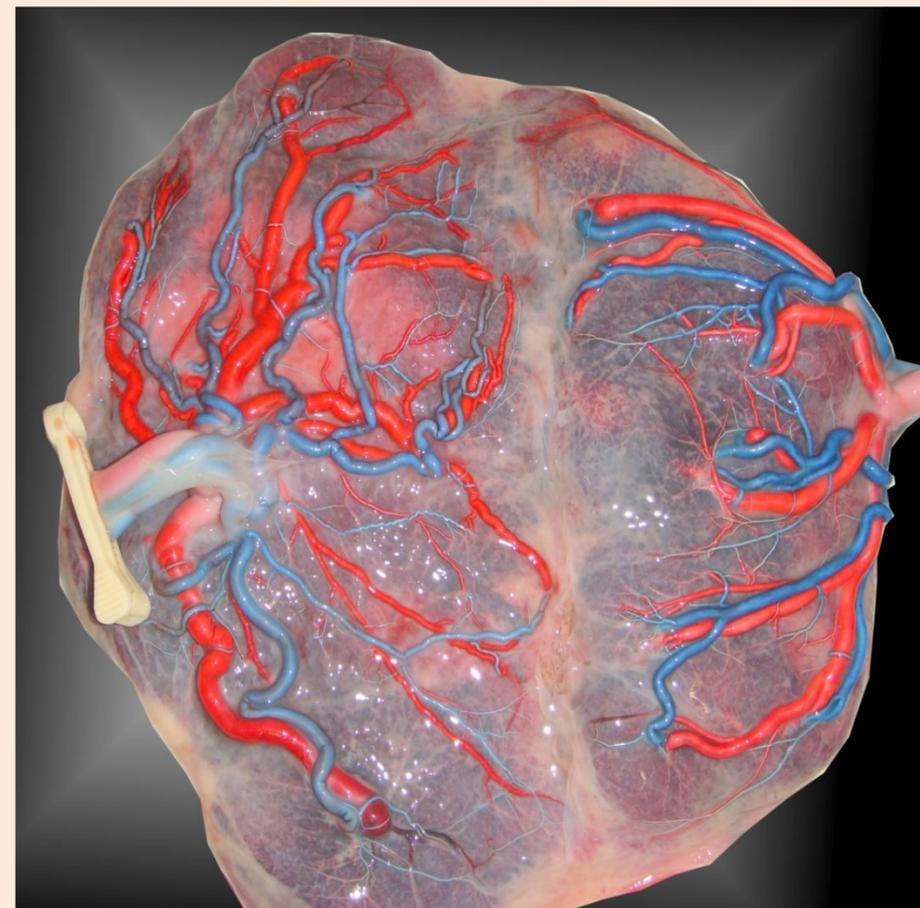
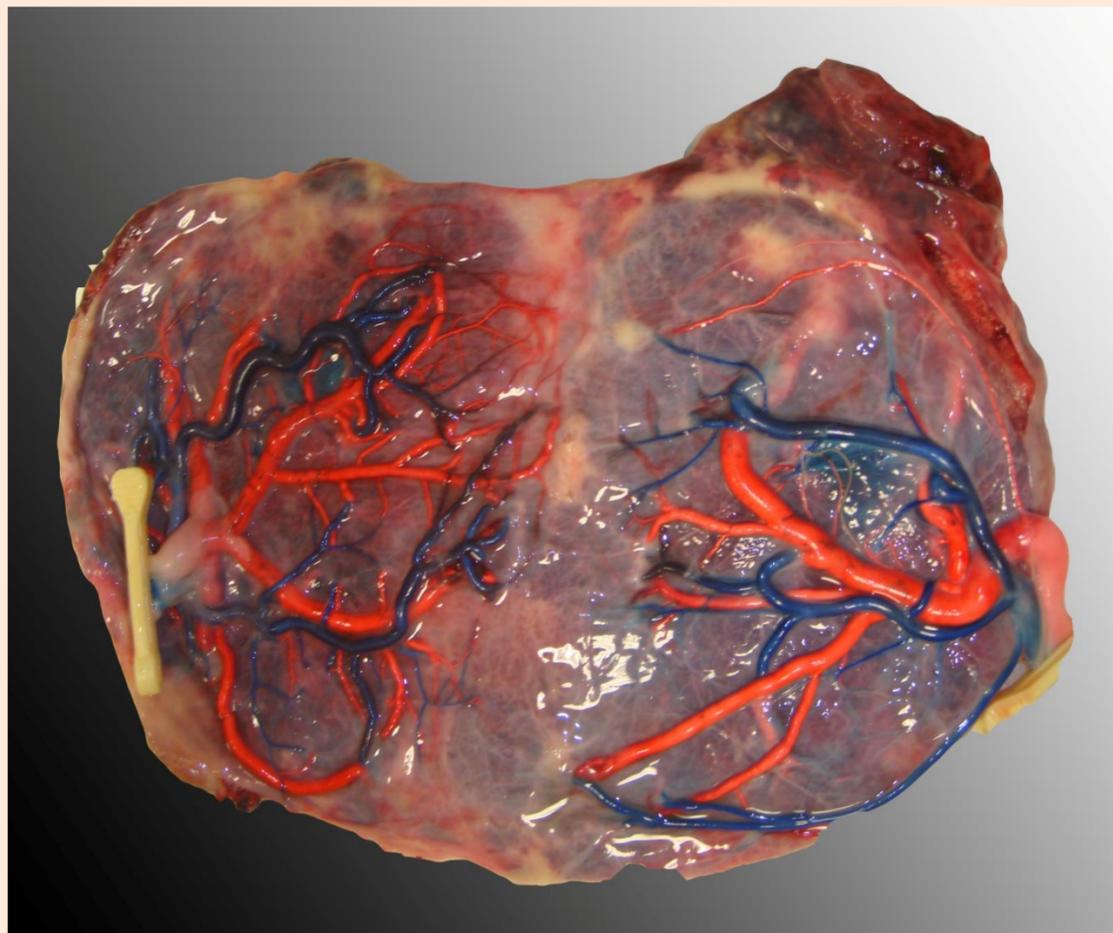
CONDITION FŒTALE:

- Discordance/RCIU
- Hémodynamique (Doppler)



Après foetoscopie laser

- Architecture placentaire après traitement laser des complications
- Important feedback pour le chirurgien
- Anastomoses résiduelles 5-32% petites et situées en périphérie
- Selon nombre, taille, type and direction: persistent STT, reversal STT, TAPS ou MIU

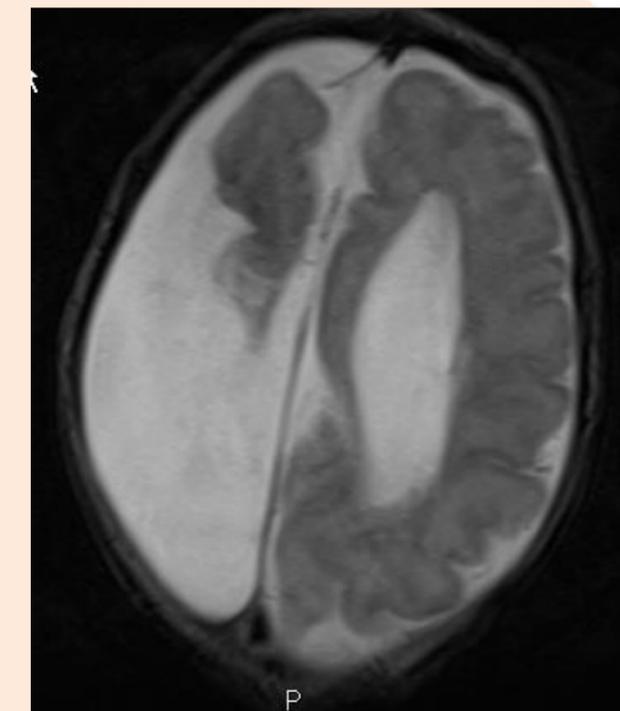
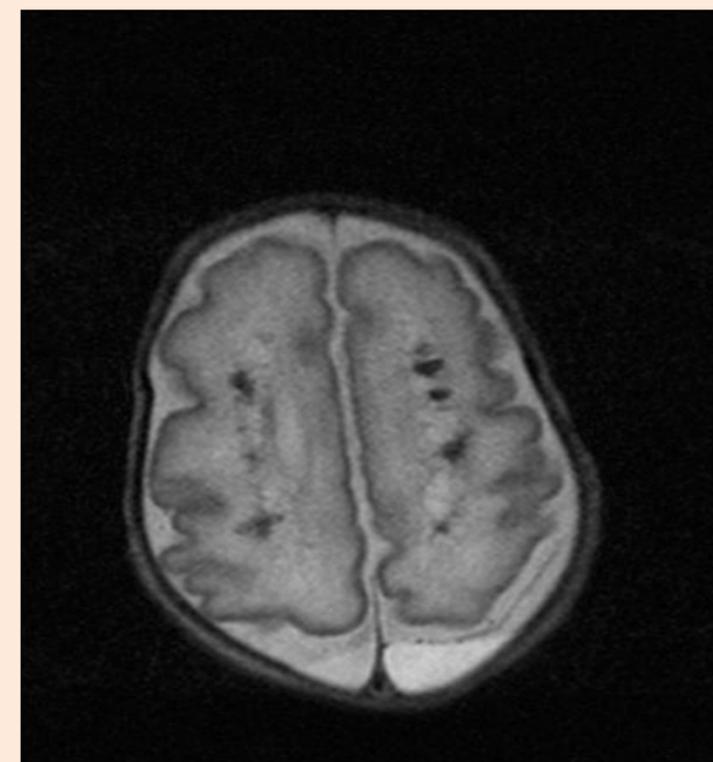
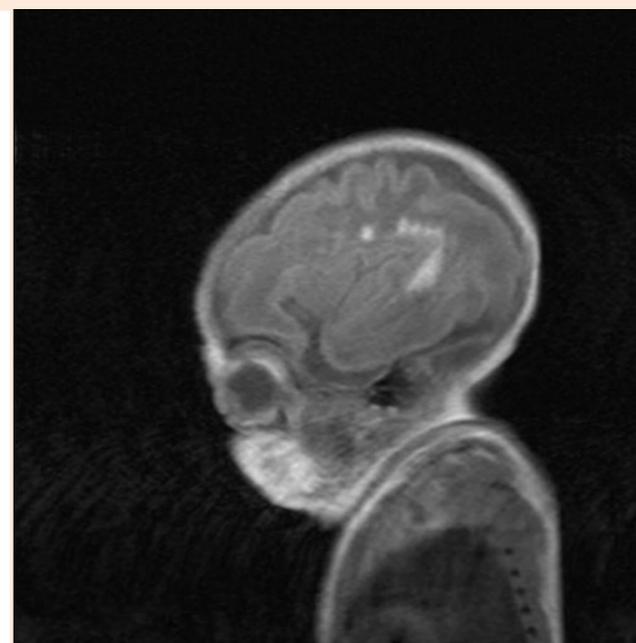


Pronostic neurologique après traitement

TRAITEMENT

| HOW DO PRETERM TWINS BORN AFTER SUCCESSFUL LASER COMPARE WITH PRETERM DICHORIONIC TWINS ? Lencien AJOG 2007 | | | |
|--|----------------------|--------------------------------|------|
| | TTTS LASER N = 97 | DICHORIONIC TWINS N = 97 | P |
| IVH 3-4 | 7.4% | 7.4% | 0.80 |
| PVL | 9.5% | 3.2% | 0.14 |
| Severe cerebral lesions (IVH or PVL) | 18% | 13% | 0.46 |
| Death or severe cerebral lesion | 21% | 18% | 0.71 |

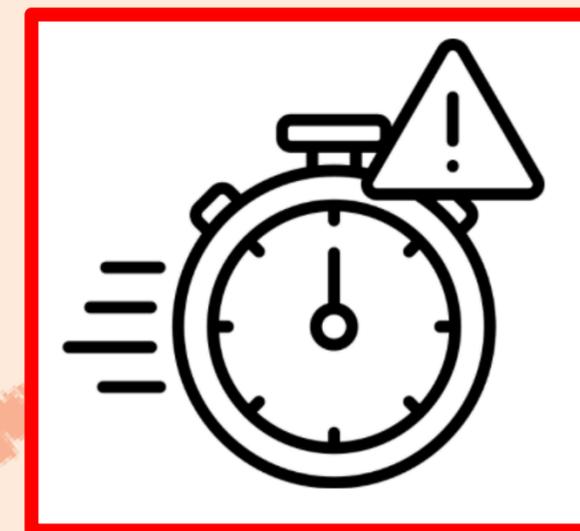
Une IRM sera systématiquement effectuée à 32 semaines car le risque résiduel de lésions neuro-encéphaliques chez ces foetus est d'environ 3%.



La plupart = neurodvpt **NORMAL**

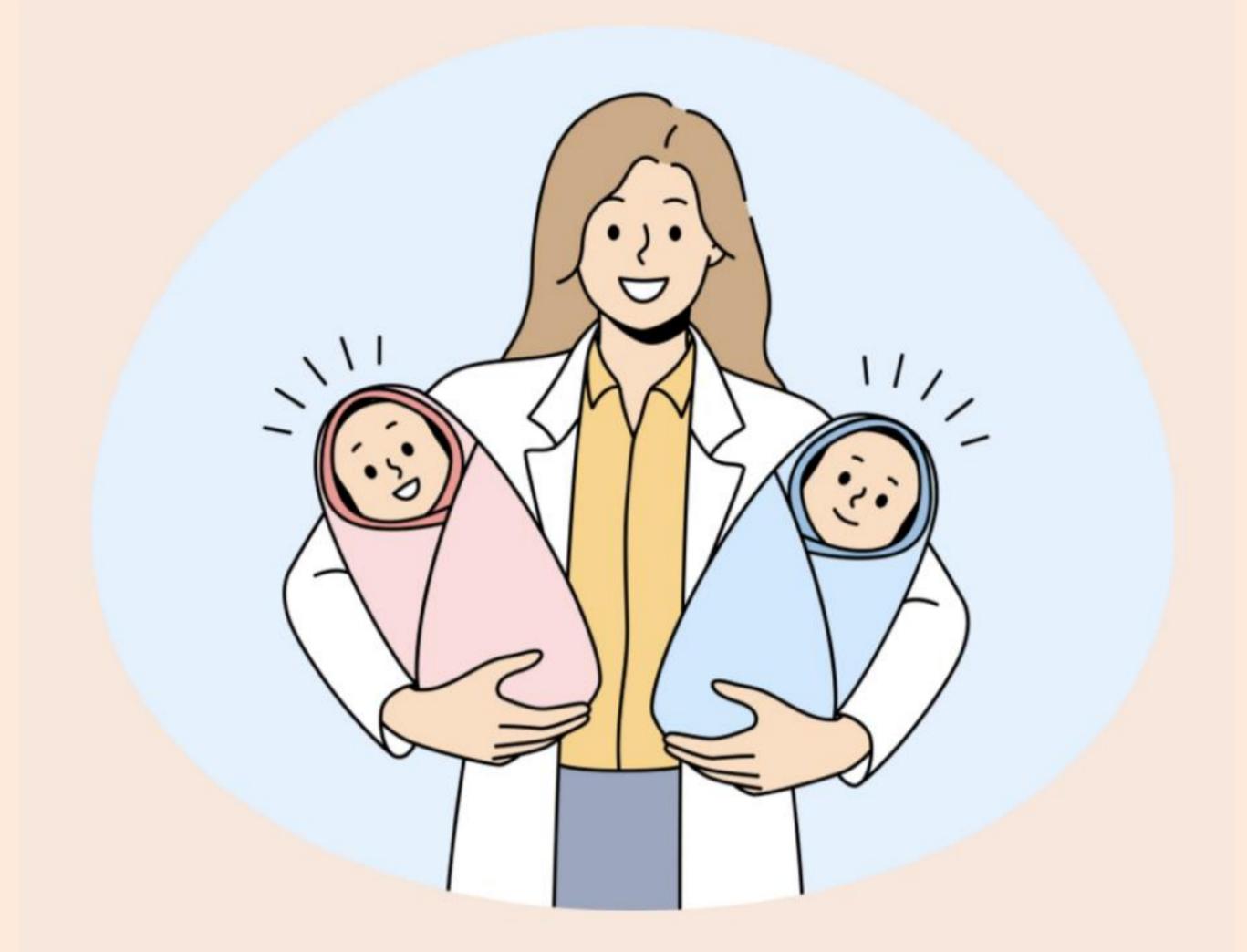
4-18% de lésions cérébrales à 2 ans malgré très bon pronostic

Dépend de la prématurité et stade initial au ttt



Adressage précoce dès suspicion

Suivi et accouchement



IRM cérébrale à 32 SA : proposée systématiquement

Accouchement en **niveau III ou adapté : ≤ 36 SA**

Suivi neuro pédiatrique ?





Protocole National de Diagnostic et de Soins (PNDS) Syndrome Transfuseur Transfusé

*Centre de Référence Pathologies rares liées au placenta des grossesses
monochoriales*

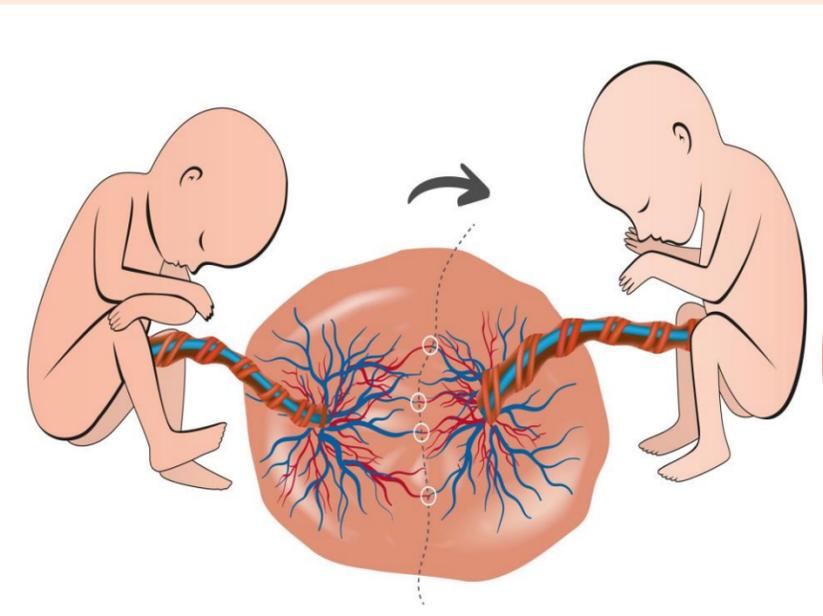
Filière AnDDI-Rares

Texte du PNDS
Décembre 2024

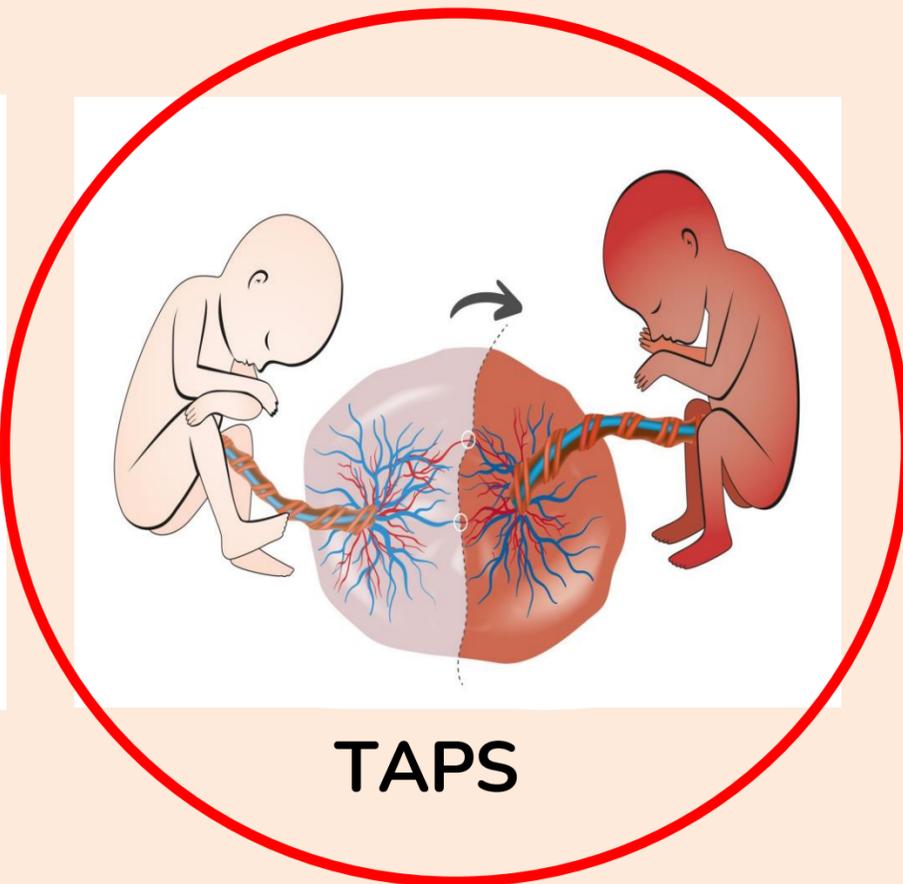
PEC thérapeutique des complications monochoriales



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Marie ZILLIOX - JAMET

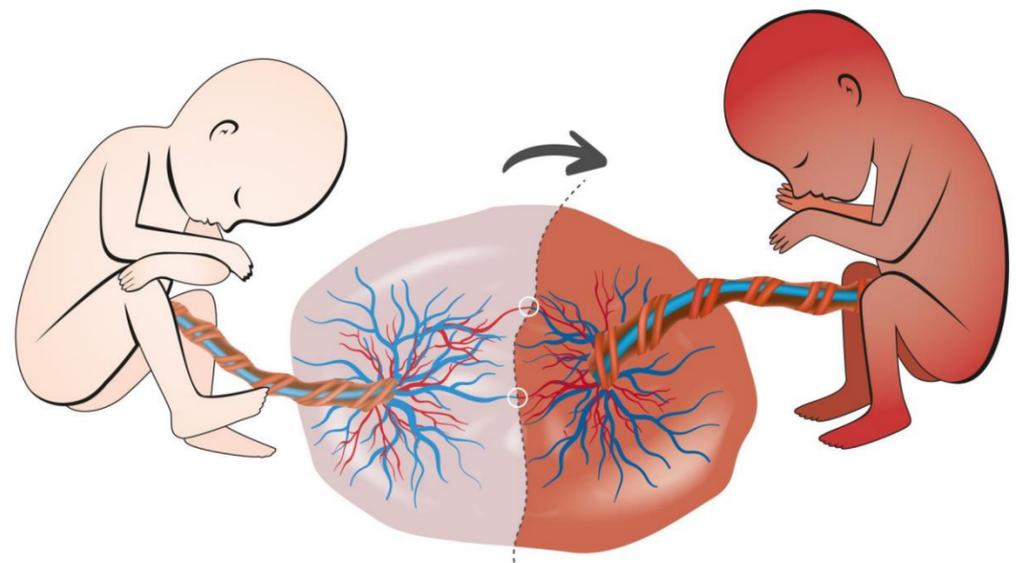


STT



TAPS

| | Solomon group (n=137 women, 274 fetuses) | Standard group (n=135 women, 270 fetuses) | OR (95% CI) |
|---|--|---|--------------------|
| Primary outcome*† | 94/274 (34%) | 133/270 (49%) | 0.54 (0.35–0.82) |
| 1-month postnatal survival | | | |
| Overall* | 203/274 (74%) | 198/270 (73%) | 1.04 (0.66–1.63) |
| At least one surviving neonate | 116/137 (85%) | 117/135 (87%) | 0.85 (0.43–1.68) |
| Double survival | 87/137 (64%) | 81/135 (60%) | 1.16 (0.71–1.89) |
| TAPS or recurrent TTTS | 6/137 (4%) | 29/135 (21%) | 0.17 (0.07–0.42) |
| Recurrent TTTS | 2/137 (1%) | 9/135 (7%) | 0.21 (0.04–0.98) |
| TAPS | 4/137 (3%) | 21/135 (16%) | 0.16 (0.05–0.49) |
| Severe neonatal morbidity‡ | 18/214 (8%) | 28/211 (13%) | 0.65 (0.31–1.36) |
| Chronic lung disease | 4/214 (2%) | 9/211 (4%) | 0.48 (0.11–2.03) |
| Patent ductus arteriosus | 6/214 (3%) | 15/211 (7%) | 0.37 (0.10–1.35) |
| Necrotising enterocolitis | 2/214 (1%) | 2/211 (1%) | 0.99 (0.14–6.97) |
| Retinopathy of prematurity§ | 0/214 (0%) | 3/211 (1%) | 7.20 (0.69–970.52) |
| Ischaemic limb injury¶ | 1/214 (<1%) | 1/211 (<1%) | 0.99 (0.06–15.72) |
| Amniotic band syndrome | 1/214 (<1%) | 3/211 (1%) | 0.33 (0.03–3.10) |
| Severe cerebral injury* | 8/169 (5%) | 10/178 (6%) | 0.85 (0.32–2.29) |
| Intraventricular haemorrhage ≥grade 3§ | 5/169 (3%) | 5/178 (3%) | 1.06 (0.27–4.10) |
| Ventricular dilatation (greater than the 97th percentile) | 1/169 (1%) | 3/178 (2%) | 0.35 (0.04–3.31) |
| Cystic periventricular leukomalacia ≥grade 2§ | 0/169 (0%) | 1/178 (1%) | 2.86 (0.15–419.23) |
| Porencephalic or parenchymal cysts§ | 1/169 (1%) | 0/178 (0%) | 0.31 (0.002–5.94) |
| Arterial infarction§ | 1/169 (1%) | 0/178 (0%) | 0.31 (0.002–5.94) |
| Other severe cerebral injury§ | 0/169 (0%) | 1/178 (1%) | 2.86 (0.15–419.23) |



Données limitées au sujet de l'impact sur le neurodéveloppement à long terme

Slaghekke et al. 2014

Après foetoscopie laser



| | | |
|----|--|---------------|
| 8 | After laser therapy, we suggest weekly surveillance for 6 weeks followed by resumption of every-other-week surveillance thereafter, unless concern exists for post-laser TTTS, post-laser TAPS, or FGR. | 2C |
| 9 | Following the resolution of TTTS after fetoscopic laser surgery and without other indications for earlier delivery, we recommend delivery of dual-surviving MCDA twins at 34 to 36 weeks of gestation. | 1C |
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Après foetoscopie laser



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Δ MCA-PSV, difference between donor and recipient MCA-PSV MoM values; FGR, fetal growth restriction; MC, monochorionic; MCDA, monochorionic-diamniotic; MCA-PSV, middle cerebral artery Doppler peak systolic velocity; MoM, multiples of the median; TAPS, twin anemia-polycythemia sequence; TTTS, twin-twin transfusion syndrome.

Society for Maternal-Fetal Medicine. Twin-twin transfusion syndrome and twin anemia-polycythemia sequence. *Am J Obstet Gynecol* 2024.



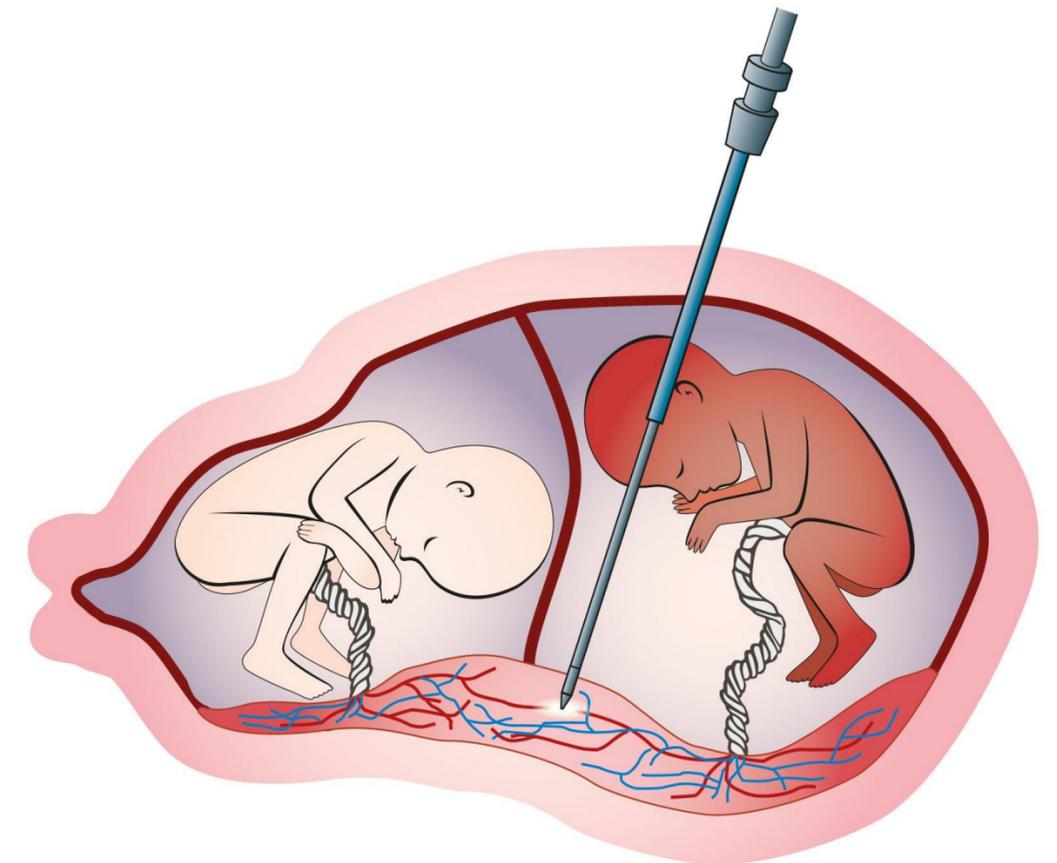
Surveillance du TAPS

- **Complication grave +++ de monochoriale**
- **Dès diagnostic, intensifier surveillance**
- **Adresser en centre de référence**

TERME

2^e et 3^e T

Moyenne 23,7 SA



Traitement

Quand? Qui?

TERME

Moyenne 23,7 SA

STADE/GRAVITE

Stade 1 tardif ? Stade 2 « précoce »?

Fact. Obstétricaux, RCIU
associé comorbidités...



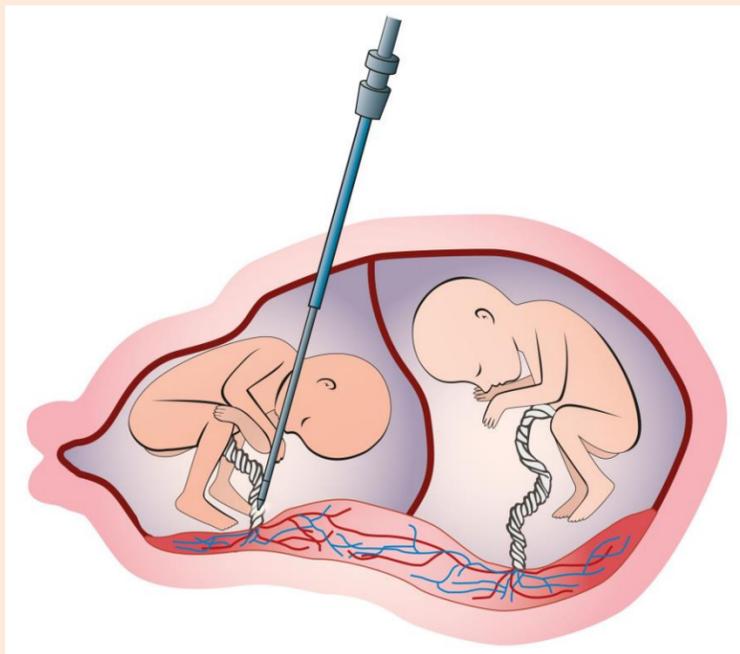
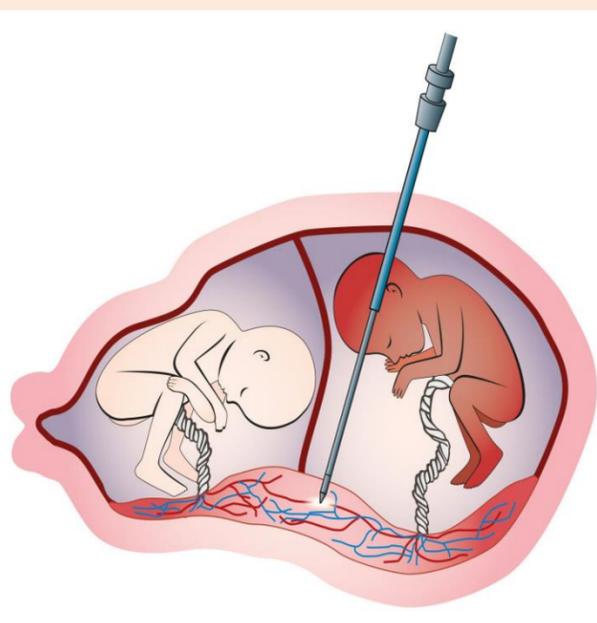
Comment?

1. **Expectative** on tempore
2. **Accouchement**
Traitement symptomatique
3. **Foetoscopie laser**
= seul traitement étiologique
4. **Transfusion in utero du donneur**
traitement symptomatique
5. **Occlusion de cordon ombilical :**
ISG

Traitement

**Stades précoces /et
sévères**

**Thérapeutique
Foetale**



**Stades tardifs et
débutants**



**Accouchement
Corticothérapie foetale**

Actuellement pas de donnée suffisante

**Essai randomisé en cours
laser/TIU/accouchement**



TAKE HOME MESSAGE

STT & TAPS



- **Monochoriales** = Gémellaire à risque spécifique
 - **STT fréquent , TAPS**
 - **Adresser dès suspicion diagnostique en centre de référence**
 - **Prise en charge thérapeutique codifiée STT , moins pour TAPS**
- Dans tous les cas ... **INDIVIDUALISEE**
- **Prise en charge pluridisciplinaire**