

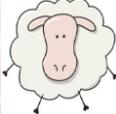
Anesthésie chez la femme enceinte en dehors de l'accouchement:
point de vue du chirurgien et de l'anesthésiste

Conséquences de la chirurgie fœtale (STT, drainages fœtaux, plugs)

Principales interventions pratiquées *in utero*

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De l'aiguille à la foetoscopie...

- Progrès du diagnostic anténatal
- Apparition de nouvelles technologies
- Utilisation de modèles animaux 

- 1961 : transfusion intra-abdominale (Liley, 1963)
- 80': chirurgie fœtale moderne
 - Michael Harrison _Université de Californie, San Francisco
 - CDH
- 1990: foetoscopie
↓ (Rodeck, King's College Hospital, London)

Amérique du Nord	Europe de l'Ouest
Chirurgie fœtale ouverte	Foetoscopie



Critères de la chirurgie foetale

- Permettre la survie du fœtus
- Au moins, prévenir les dommages permanents
 - Par correction anatomique de la malformation
 - Par arrêt de l'évolution de la pathologie (pour une réparation PN)

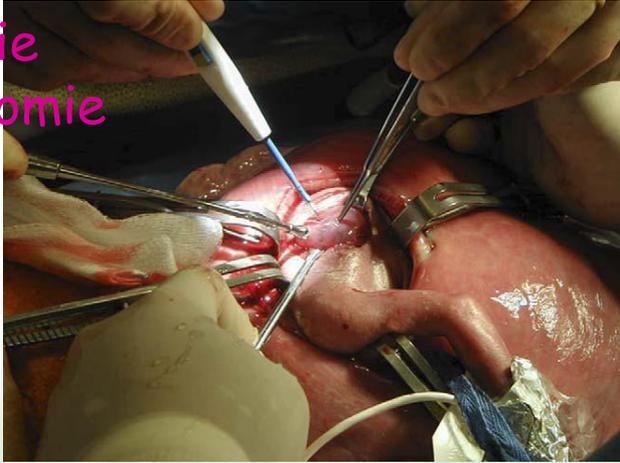
1. Accurate diagnosis and staging possible, with exclusion of associated anomalies.
2. Natural history of the disease is documented, and prognosis established.
3. Currently no effective postnatal therapy.
4. In-utero surgery proven feasible in animal models, reversing deleterious effects of the condition.
5. Interventions performed in specialized multidisciplinary fetal treatment centers within strict protocols and approval of the local ethics committee with informed consent of the mother or parents.

Harrison, 1982

Voies d'abord

Chirurgie fœtale ouverte

Laparotomie
+ hystérotomie



Laparotomie
+ endoscopie

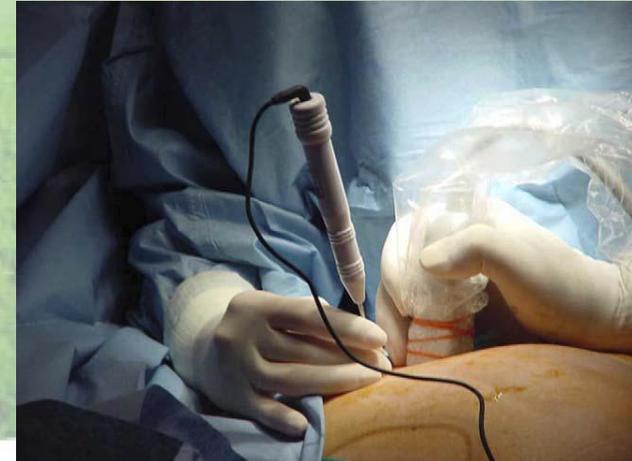


Chirurgie fœtale mini-invasive

Foetoscopie



Echoguidée



Golombek, 2006

Indications en chirurgie foetale

Fetal surgery	Pathophysiology	Rationale for in-utero therapy
1. CDH	Pulmonary hypoplasia and PHT	Reversal of pulmonary hypoplasia and prevent PHT
2. LUTO	Progressive renal damage by obstruction Pulmonary hypoplasia by oligohydramnios	Urinary diversion prevents obstructive uropathy and spares renal function Urinary diversion restores amniotic fluid
3. SCT	High-output cardiac failure by arteriovenous shunting Fetal anemia by tumor growth and/or bleeding within a tumor	Cessation of steal phenomenon Reversal of cardiac failure Prevent polyhydramnios
4. Thoracic space occupying lesions	Pulmonary hypoplasia (space-occupying mass); hydrops by impaired venous return (mediastinal compression)	Prevention of pulmonary hypoplasia and cardiac failure
5. Neural tube defects	Damage to exposed neural tube Cerebrospinal fluid leak, leading to Chiari malformation and hydrocephalus	Covering exposed spinal cord, cessation of leakage preventing hydrocephaly and reversing cerebellar herniation
6. Cardiac malformations	Critical lesions causing irreversible hypoplasia or damage	Prevention of hypoplasia or arrest of progression of damage
Surgery on the placenta, cord or membranes		
7. Chorioangioma	High-output cardiac failure by arteriovenous shunting and polyhydramnios	Prevention of cardiac failure and hydrops fetoplacentalis
8. Amniotic bands	Progressive constrictions causing irreversible neurological or vascular damage	Prevention of limb deformities and function loss
9. Abnormal monochorionic twinning: twin-to-twin transfusion	Intertwin transfusion leads to oligo-polyhydramnios sequence, hemodynamic changes; obstetrical complications (preterm labor and rupture of the membranes)	Bichorionization stops intertwin transfusion, reverses cardiac failure Preventing neurological damage Delaying delivery (amniodrainage)
Fetus acardiacus and discordant anomalies	Discordant anomalies: where one fetus can be a threat to the other one or to avoid termination of entire pregnancy	Feticide to improve chances of the other fetus; avoidance of termination of entire pregnancy

CDH, congenital diaphragmatic hernia; LUTO, lower urinary tract obstruction; PHT, pulmonary hypertension; SCT, Sacrococcygeal teratoma.

Deprest, 2010

45^{ème} congrès du CARO, Clermont-Ferrand, 11/12 Mai 2012

Hernie diaphragmatique congénitale

Pathophysiology

Rationale for in-utero therapy

Pulmonary hypoplasia and PHT

Reversal of pulmonary hypoplasia and prevent PHT

- **Réparation prénatale du défaut** (Harrison, 1997)

- **OT:**

✓ **Clip:** laparotomie, hystérotomie (Flake, 2000) ou endoscopie

(Harrison, 2003)

✓ **OT par ballonnet:**

• laparotomie, endoscopie (Harrison, 2001)

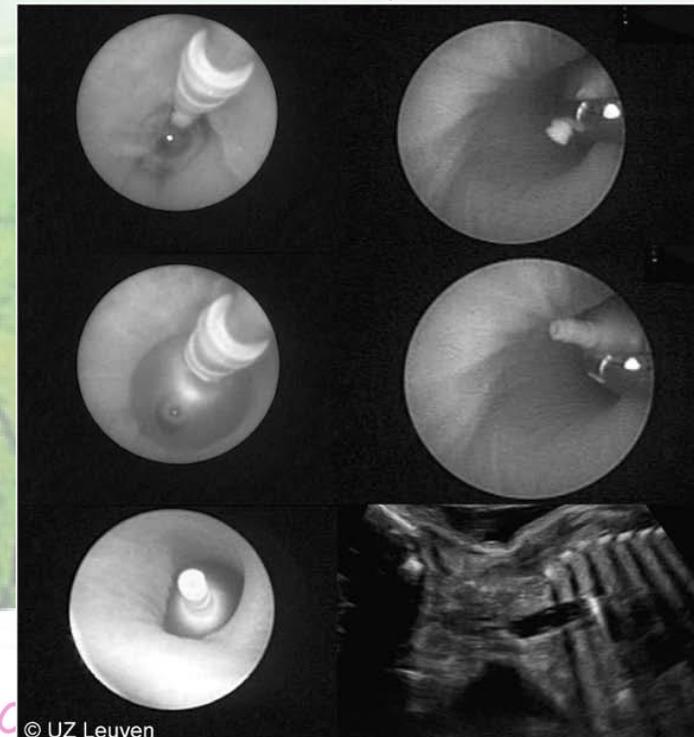
• foetoscopie (Quintero, 2000)

FETO (Deprest, Gratacos, Nicolaides)

- retrait par EXIT (Bouchard, 2002)

- retrait par ponction échoguidée

ou par foetoscopie



Hernie diaphragmatique congénitale

The making of fetal surgery

Jan A. Deprest^{1,2*}, Alan W. Flake³, Eduard Gratacos^{2,4}, Yves Ville^{2,5}, Kurt Hecher^{2,6}, Kypros Nicolaides^{2,7}, Mark P. Johnson³, François I. Luks⁸, N. Scott Adzick³ and Michael R. Harrison⁹

Prenat Diagn 2010; **30**: 653–667.

	Harrison <i>et al.</i> (2003)	FETO consortium (2009)
Criteria for surgery	LHR < 1.4 and liver 'up'	LHR < 1.0 and liver 'up'
Anaesthesia	General	Loco-regional or local
Access through abdominal wall	Laparotomy	Percutaneous
Access diameter	5 mm cannula	3.3 mm cannula
Occlusive device	Clip or endoluminal balloon	Endoluminal balloon
Reversal of occlusion	EXIT delivery	<i>In utero</i> reversal
PPROM < 34 weeks	100%	25%
Mean gestational age at birth	30.8 (28–34)	35.3 weeks (25.7–41.0)
Survival following TO (LHR < 1.4)	73% (<i>n</i> = 11) (controls: 77%)	TO not performed in this group
Survival following TO (LHR < 1.0)	33% (<i>n</i> = 3) (left CDH)	Left-CDH: 49% Right-CDH: 35%

HDC G sévère: 24,1%
HDC D sévère 0%
(Jani, 2006)

Uropathies obstructives (LUTO: Lower Urinary Tract Obstruction)

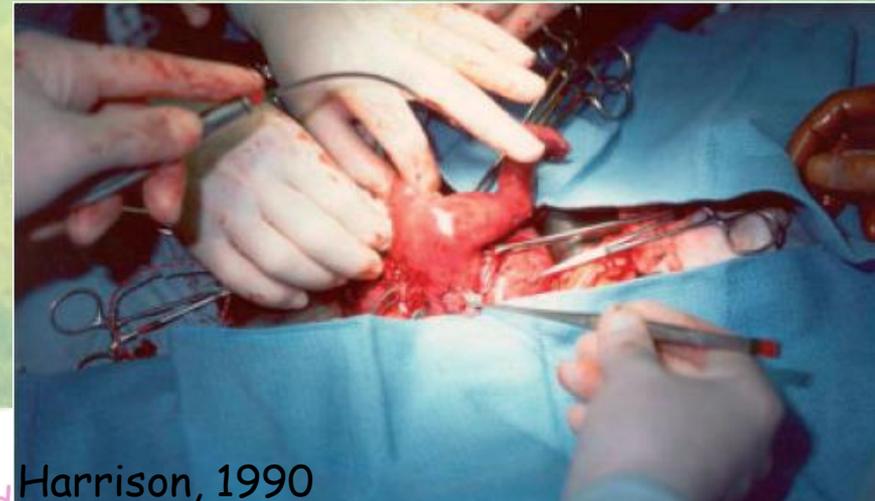
Pathophysiology

Progressive renal damage by obstruction
Pulmonary hypoplasia by oligohydramnios

Rationale for in-utero therapy

Urinary diversion prevents obstructive uropathy and spares renal function
Urinary diversion restores amniotic fluid

- Mortalité périnatale > 90%
- Insuffisance rénale > 50% (Parkhouse, 1988; Freedman, 1999)
- Thérapies fœtales:
 - **Urétérostomies fœtales par hystérotomie** (Harrison, 1981): abandonnée
 - **Vésicocentèses** (répétées)



Harrison, 1990

Uropathies obstructives

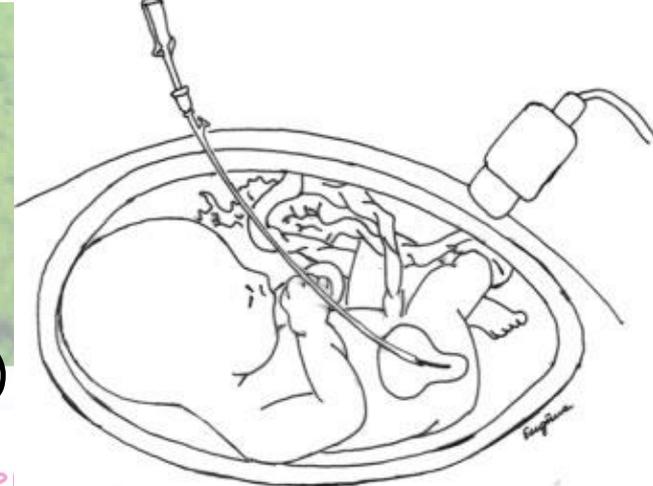
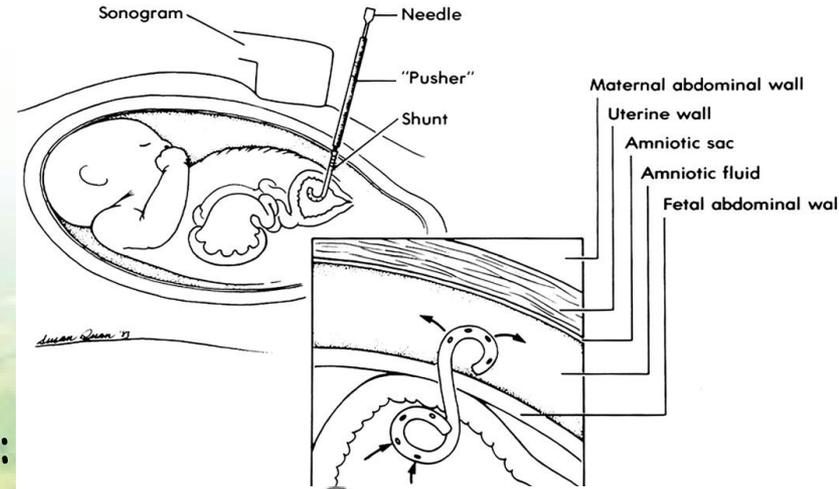
Thérapies fœtales:

- Drain vésico-amniotique sous échoguidage

- 45% de complications: obstruction, migration, RPM, AP, chorio,
- 40% de survie, 50% d'IR (Holmes, 2001)

- Cystoscopie fœtale (Quintero, 1995):

- diagnostic de VUP (95%),
- ttt (hydro-ablation, laser, cathérisation antérograde)
→ drainage physiologique
- 70-75% de survie néonatale (Ruano, 2011)



Tératome sacro-coccygien

Pathophysiology	Rationale for in-utero therapy
High-output cardiac failure by arteriovenous shunting Fetal anemia by tumor growth and/or bleeding within a tumor	Cessation of steal phenomenon Reversal of cardiac failure Prevent polyhydramnios

- Morbidité fonction de l'extension
 - anémie,
 - fistule AV → IC → anasarque, hydramnios,
 - MFIU (25% (Benachi, 2006))
- >7 cm / SF
 - Résection par hystérotomie (Hedrick, 2004)
 - Coag des vx par laser, thermocoag interstitielle, radiofq (Lam, 2002)
 - Embolisation: alcool (Makin, 2007), histoacryl (Perrotin, 2006), coils

Pathologies thoraciques

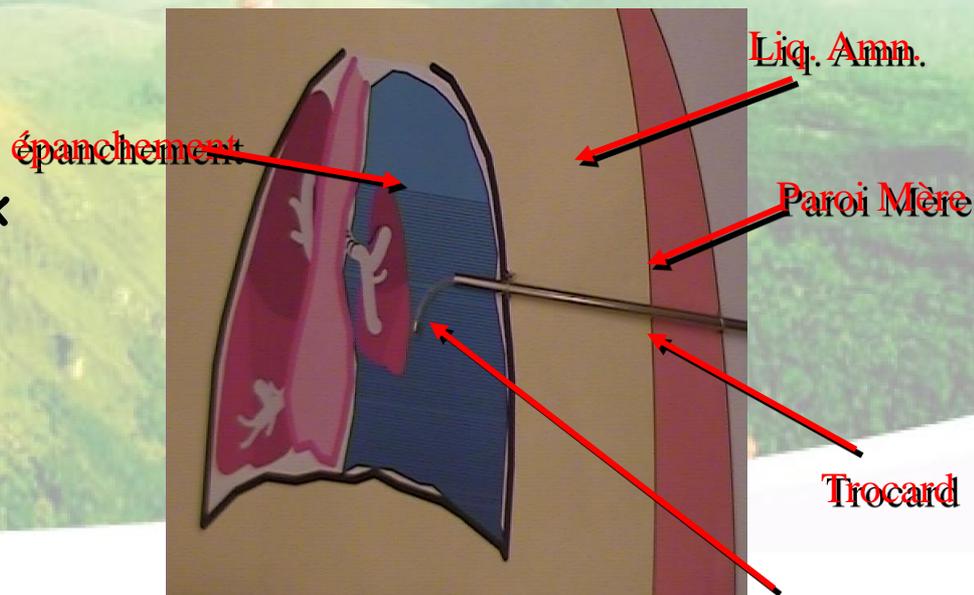
Pathophysiology

Rationale for in-utero therapy

Pulmonary hypoplasia (space-occupying mass); hydrops by impaired venous return (mediastinal compression)

Prevention of pulmonary hypoplasia and cardiac failure

- **Malformations adénomatoïdes kystiques pulmonaires (MAKP)**
 - Malformation des bronchioles terminales
 - microkystique: **lobectomie fœtale**, 50% de survie (Adzick, 2010)
 - macrokystique: **ponction/drain thoraco-amniotique**, 65-75% de survie (Mann, 2010)
- **Épanchement pleural - Chylothorax**
 - **Ponction ou drain**
 - 70% de survie (Mann, 2010)



Myéломéningocèle

Pathophysiology

Damage to exposed neural tube
Cerebrospinal fluid leak, leading to Chiari malformation
and hydrocephalus

- Spina bifida ouvert
- troubles moteurs et cognitifs
- Ttt PN: 85% shunt ventriculopéritonéal,
- 15% de DC à 5 ans
- Rationnel du ttt *in utero*:
 - arrêt de la fuite de LCR et
 - arrêt de l'exposition au LA

Rationale for in-utero therapy

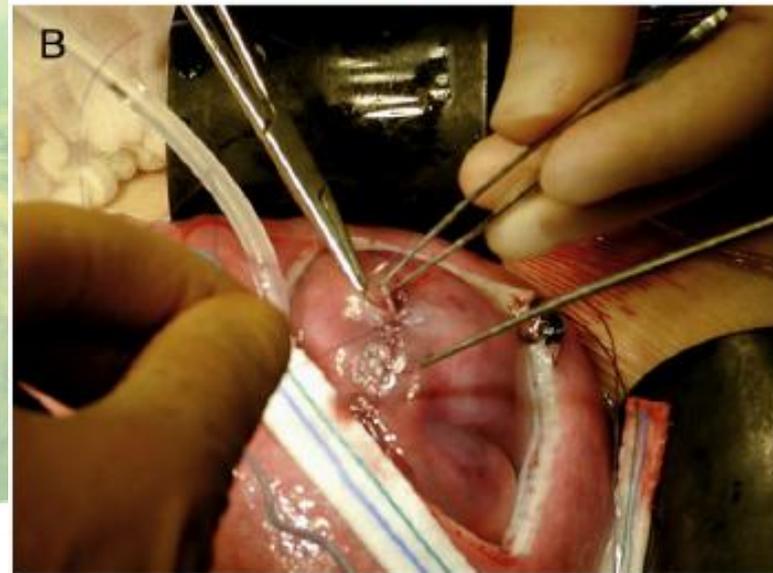
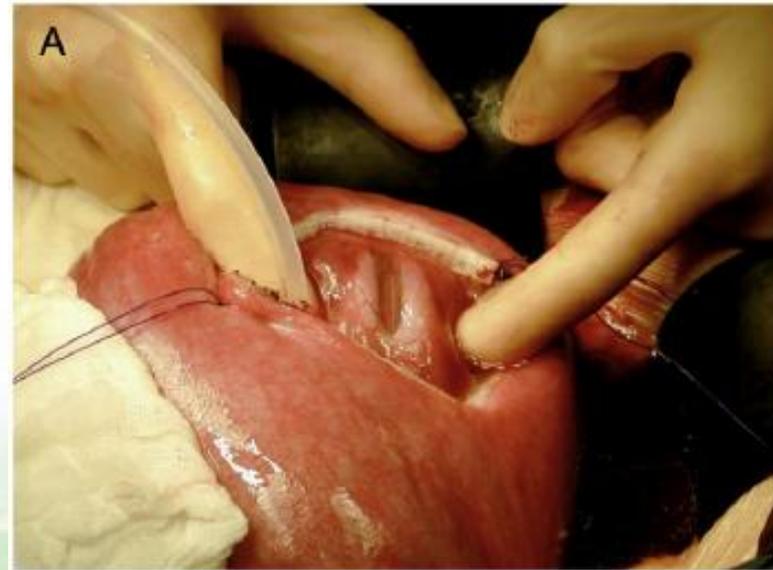
Covering exposed spinal cord, cessation
of leakage preventing hydrocephaly
and reversing cerebellar herniation



Adzick, 2012

Myéломéningocèle

- **Fermeture du défaut** (Adzick, Lancet, 1998)
 - 40% shunt VP (vs. 82% en PN),
42% de marche (vs. 21%),
36% correction hernie FCP (vs. 4%)
(Adzick, 2011),
83% d'enfant avec fonctions
cognitives normales (Danzer, 2010)
- **Couverture par patch per foetoscopie**
(Europe) (Verbeek, 2010)
 - 69% de couverture complète,
 - 81% de survie
 - + d'AP, RPM, chorio,
 - persistance hernie FCP



Chorioangiome

Pathophysiology

High-output cardiac failure by arteriovenous shunting and polyhydramnios

- Vol vasculaire
- insuffisance cardiaque, anasarque, hydramnios (18-35%),
- TTT: **dévascularisation per foetoscopie ou embolisation** (Nicolini, 1999; Quarello, 2005)

Rationale for in-utero therapy

Prevention of cardiac failure and hydrops fetoplacentalis

Maladie des brides amniotiques

Pathophysiology

Progressive constrictions causing irreversible neurological or vascular damage

- amputation de membres
- TTT: **section per foetoscopie**
(Quintero, 1997)



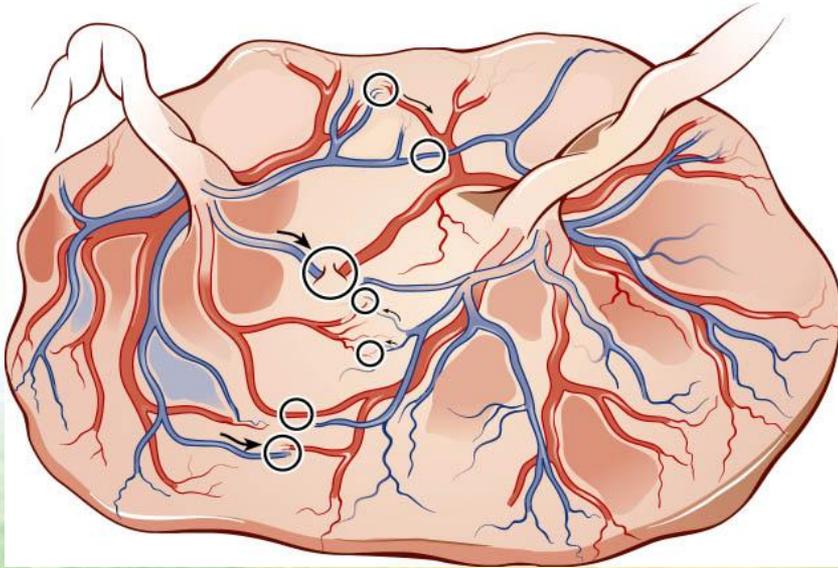
Rationale for in-utero therapy

Prevention of limb deformities and function loss

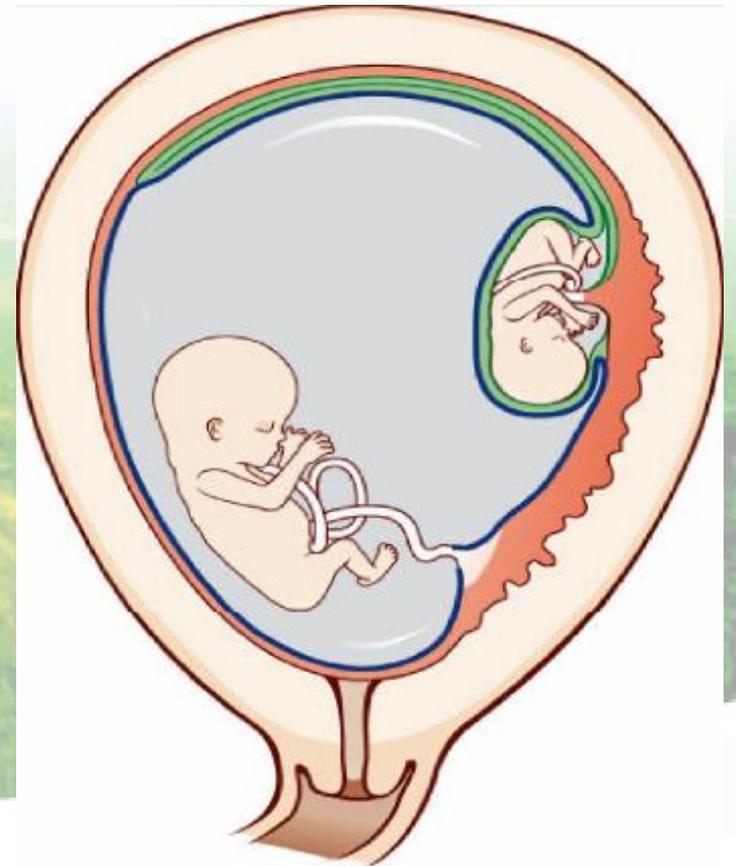


© Fetal Care Center of Cincinnati, 2007

Traitements *in utero* des complications des grossesses gémellaires monochoriales



- **Syndrôme transfuseur-transfusé**
 - séquence oligoamnios-hydramnios
 - 10-15% des GG monochoriales
 - 80-100% de décès < 26SA



Traitements *in utero* des complications des grossesses gémellaires monochoriales

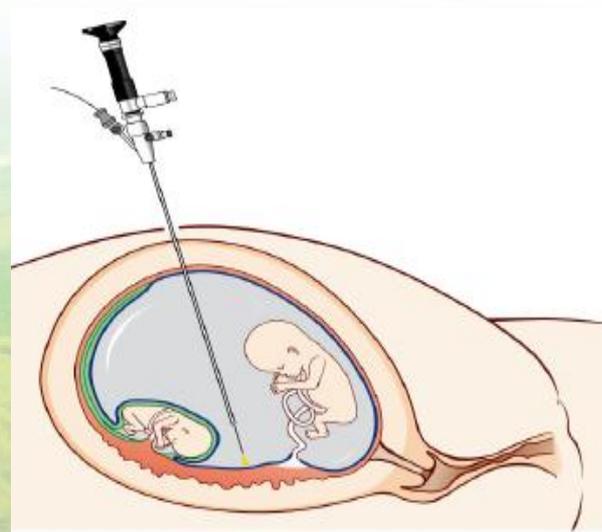
Pathophysiology

Intertwin transfusion leads to oligo-polyhydramnios sequence, hemodynamic changes; obstetrical complications (preterm labor and rupture of the membranes)

Rationale for in-utero therapy

Bichorionization stops intertwin transfusion, reverses cardiac failure
Preventing neurological damage
Delaying delivery (amniodrainage)

- **Syndrôme transfuseur-transfusé**
 - Drainage amniotique
 - Photocoagulation-laser
- **Autres indications de la PL**
 - TAPS
 - RCIU sélectif



	PL (%)		AI (%)	
	Rossi et D'Addario [4]	Notre étude	Rossi et D'Addario [4]	Notre étude
Taux de survie à la naissance	57-77	75	38-81	53
Aucun survivant	13-25	12	9-49	32
Au moins un survivant	75-87	88	51-90	68
Décès néonatal avant 28 jours de vie	4-12	4	14-39	5
Anomalies cérébrales après 28 jours de vie	2-33	3	18-83	10

Traitements *in utero* des complications des grossesses gémellaires monochoriales

Pathophysiology

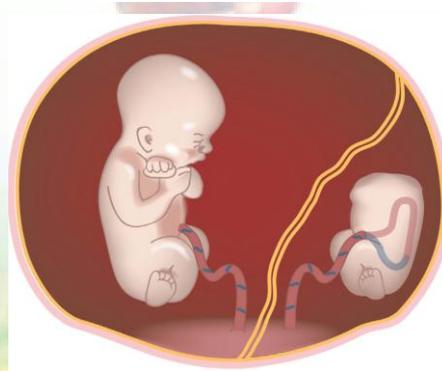
Discordant anomalies: where one fetus can be a threat to the other one or to avoid termination of entire pregnancy

• Foeticide sélectif

- Indications

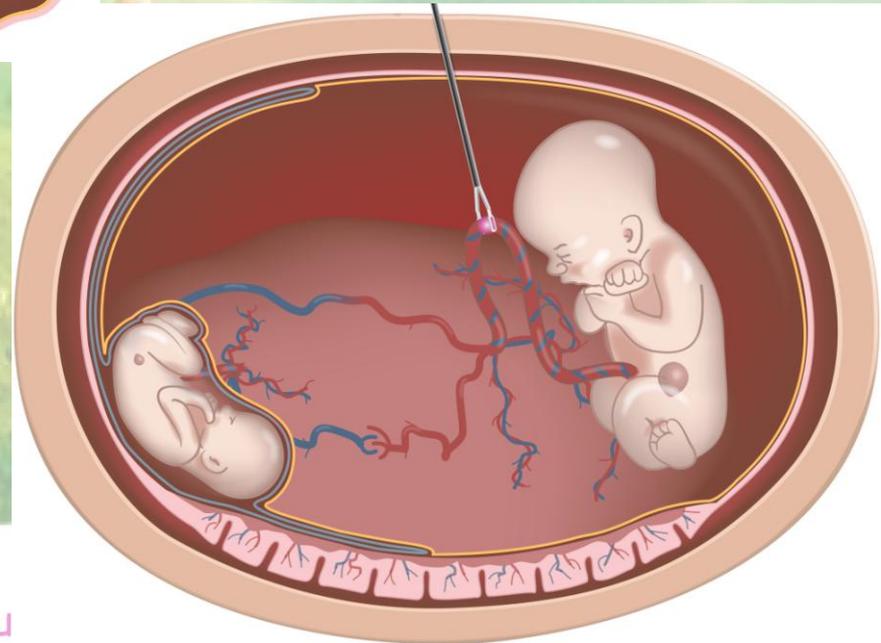
- STT, sRCIU
- TRAP
- Malformation

- #### - Techniques: Occlusion cordonale par pince bipolaire, laser, radiofréquence



Rationale for in-utero therapy

Feticide to improve chances of the other fetus; avoidance of termination of entire pregnancy



Interventions sur le cœur foetal

Pathophysiology

Critical lesions causing irreversible hypoplasia or damage

Rationale for in-utero therapy

Prevention of hypoplasia or arrest of progression of damage

- **Nouvelles thérapies foetales**
- TTT (Gardiner, 2012):
 - Valvuloplastie des valves aortique et pulmonaire
 - Septostomie par ballonnet (sténose Ao, hypoVG, TGV)
 - Stimulation pour BAV (Carpenter, 1986; Assad, 2003)
- Techniques:
 - Accès percutané
 - Echoguidage
 - Ponction cardiaque



EXIT

Ex-Utero Intrapartum Treatment

Indications for an EXIT Procedure

EXIT-to-airway procedure

Neck mass with obstruction (eg, cervical teratoma, vascular/lymphatic malformation, goiter, neuroblastoma)

Congenital high airway obstruction syndrome

Tracheal atresia

Laryngeal atresia

Micrognathia

Other causes of airway obstruction

EXIT-to-resection procedure

Chest masses with intrathoracic airway obstruction

Congenital pulmonary airway malformation

Bronchopulmonary sequestration

Mediastinal teratoma

Pericardial teratoma

EXIT-to-ECMO procedure

Severe CDH

Severe congenital heart disease

EXIT-to-separation procedure

Conjoined twins

- Intubation intra-trachéale, trachéotomie, trachéoplastie, ablation tumorale
- Retrait du ballonnet d'OT dans la hernie diaphragmatique congénitale



Figure 12. Hysterotomy and EXIT procedure. Intraoperative photographs obtained during maternal laparotomy performed using a Pfannenstiel incision to access the abdominal cavity show the fetal head being delivered through the incision (a) and a laryngoscope being passed into the fetal trachea (b). A large, multicystic neck mass was seen inferior to the endotracheal tube.