
SoFFœt
Société Française de Fœtopathologie

# Malformations Pulmonaires de Diagnostic Anténatal.

# Prise en Charge Ante-Natale.

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**Hôpital Femme Mère Enfant - Lyon**

Dr M Massoud, Pr L Guibaud, Dr M Cagneaux, Dr C Dufour

# *Généralités*


- Epidémiologie ?
- Problème de classification anténatale
  - Classifications Historiques (Stocker 1977, Adzick)
  - Classifications Histologiques ? (Langston 2003)
- ***MALFPULM :*** 3 types
  - **Lésion Echogène pure non kystique**
  - **Lésion Kystique Pure**
  - **Lésion Mixte**



Prenatal natural history of congenital pulmonary malformations: MALFPULM population-based cohort study

*Ultrasound Obstet Gynecol* 2019; **54**: 381–388

C. DELACOURT[1,2], N. BERTILLE[3], L. J. SALOMON[2,4], A. BENACHI[5], E. HENRY[6], J. MASSARDIER[7], N. MOTTET[8], J. ROSENBLATT[9], A. SARTOR[10], C. THONG-VANH[11], A. S. VALAT-RIGOT[12], N. WINER[13], N. LELONG[3] and B. KHOSHNOOD[3], for the Prenatal MALFPULM Study Group

# *Modes de découverte et Bilan*

## Echographie 2ème trimestre

**Bilan :**

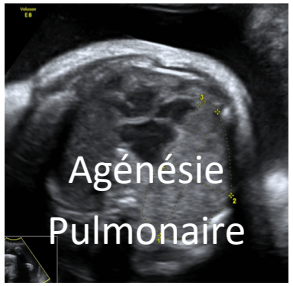
- IRM non recommandée en routine *PNDS 2020*
- Pas de caryotype/ACPA de principe
- Pas d'exploration cardiaque de principe

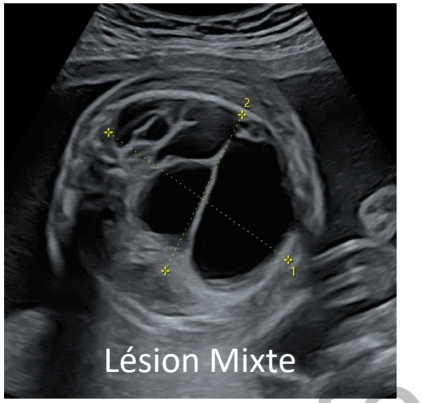
**Table 1** Comparison of study population and excluded population of pregnancies with diagnosis of fetal congenital pulmonary malformation (CPM)

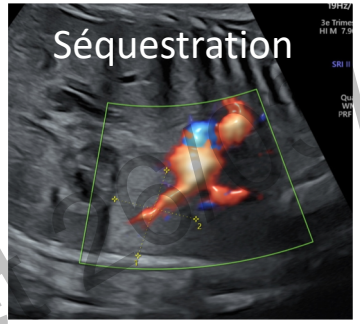
| *Characteristic* | *Total* (n = *189*) | *Study population* (n = *176*) | *Excluded* (n = *13*) | P |
|---|---|---|---|---|
| US per woman (*n*) | 4.3 ± 0.1 | 4.4 ± 0.1 | 2.2 ± 0.4 | < 0.001 |
| MA (years) | 29.9 ± 0.4 | 29.8 ± 0.4 | 30.0 ± 1.3 | 0.92 |
| GA at first US (weeks) | 24.6 ± 0.3 | 24.4 ± 0.2 | 27.1 ± 1.5 | 0.008 |
| CPM phenotype at first US | | | | 0.02 |
| Hyperechoic | 93 (49.2) | 90 (51.1) | 3 (23.1) | |
| Cystic/mixed | 96 (50.8) | 86 (48.9) | 10 (76.9) | |
| CPM-related sign of compression | | | | |
| Any | — | 80 (45.5) | — | |
| Mediastinal shift | — | 79 (44.9) | — | |
| Eversion of diaphragm | — | 20 (11.4) | — | |
| Ascites | — | 8 (4.5) | — | |
| Polyhydramnios | — | 15 (8.5) | — | |
| Hydrothorax | — | 7 (4.0) | — | |
| Hydrops | — | 3 (1.7) | — | |

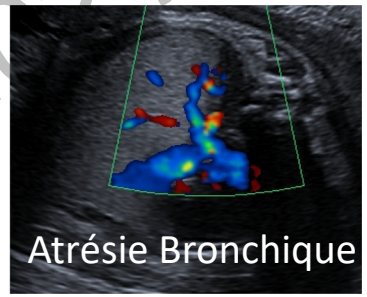
Data are given as mean ± standard error or *n* (%). GA, gestational age; MA, maternal age at inclusion; US, ultrasound examination.

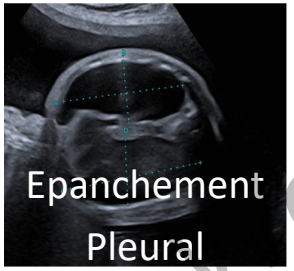
# *Type lésionnel*

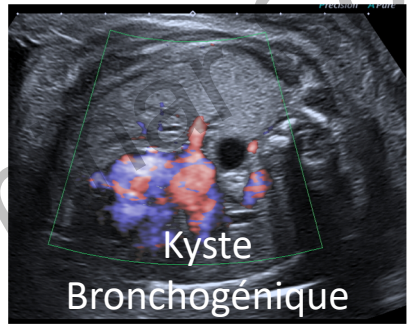


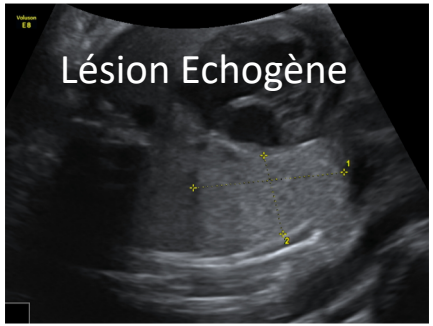




**Tout le reste :**

Emphysème

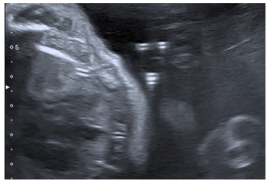
Lymphangiome

Tumeurs

CHAOS

….

# *Prise en charge thérapeutique in Utero*



**Ponction +/- Drainage**
En cas d'anasarque ou de compression, sur forme macrokystique

## Prenatal steroids for microcystic congenital cystic adenomatoid malformations

Journal of Pediatric Surgery (2010) 45, 145–150

Patrick F. Curran[a,b], Eric B. Jelin[a,b], Larry Rand[a,c], Shinjiro Hirose[a,b], Vickie A. Feldstein[a,c,d], Ruth B. Goldstein[a,c,d], Hanmin Lee[a,b,*]

**Table 3** Multicenter analysis of outcomes in fetuses with and without hydrops

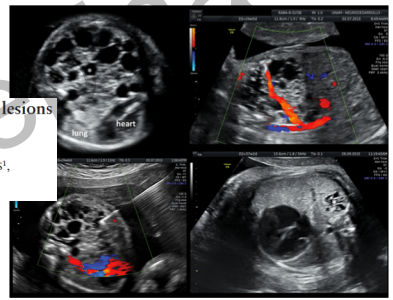
| | No hydrops | Hydrops | Hydrops, resolved | Hydrops, persistent |
|---|---|---|---|---|
| Survival | 11 (100.0%) | 16 (80.0%) | 15 * (93.8%) | 1 * (25.0%) |
| GA (delivery) (wk) | 39.0 | 36.0 | 37.0 * | 28.9 * |

| | Survival | GA (delivery) (wk) |
|---|---|---|
| No hydrops | 11 (100.0%) | 39.0 |
| Hydrops | 16 (80.0%) | 36.0 |
| Resolved | 15 * (93.8%) | 37.0 * |
| Persistent | 1 * (25.0%) | 28.9 * |

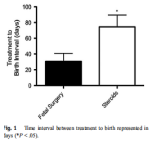
Laser interstitiel

## Fetal laser ablation of feeding artery of cystic lung lesions with systemic arterial blood supply

R. CRUZ-MARTINEZ[1,2], M. MARTÍNEZ-RODRÍGUEZ[1,2], M. BERMÚDEZ-ROJAS[1], C. MAGAÑA-ABARCA[1], V. NARVAEZ-DOMINGUEZ[2], A. ROJAS-MACEDO[3], N. BAUTISTA-GARCÍA[2] and M. ALCOCER-ALCOCER[2]




Chirurgie Ouverte ?



## Microcystic congenital pulmonary airway malformation with hydrops fetalis: steroids vs open fetal resection

Kenneth C. Loh, Eric Jelin, Shinjiro Hirose, Vickie Feldstein, Ruth Goldstein, Hanmin Lee*


## *Surveillance Prénatale*

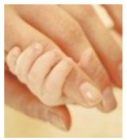
**Buts :**

- Evoquer un diagnostic
- Rechercher des malformations associées
- Rechercher des signes des gravités et organiser la naissance

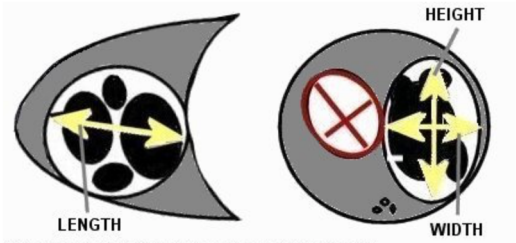
**Faire rencontrer les acteurs de la prise en charge post natale :**

- Chirurgien Pédiatre
- Pédiatre / Pneumopédiatre
- Néonatalogue

# Critères pronostics et lieu de naissance



**perinatology.com**
**The Congenital Pulmonary Airway Malformation Volume Ratio (CVR)**




**The calculator below may be used to estimate the CVR.**

Enter in centimeters:

| | |
|---|---|
| Mass Length | 1.2 |
| Mass Width | 2.4 |
| Mass Height | 4.5 |
| Fetal Head Circumference | 22.5 |

Calculate Reset

**RESULTS**

**Mass width: 2.4 cm. Height: 4.5cm . Length: 1.2 cm. Head circumference: 22.5 cm.**

**Congenital Pulmonary Airway Malformation Volume =6.74 $cm^3$**

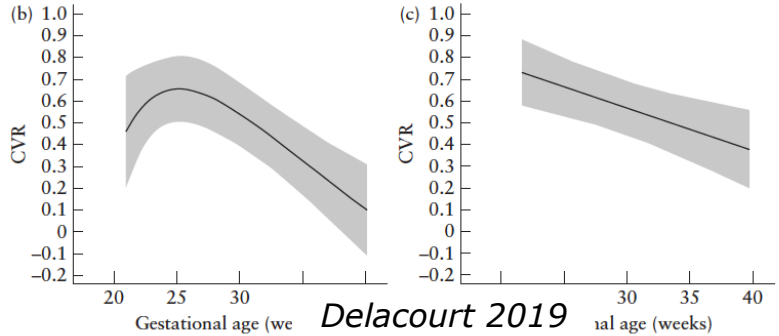
**Congenital Pulmonary Airway Malformation Volume Ratio (CVR) =0.3 $cm^2$.**

**CVR**

Prédire le risque de détresse respiratoire néonatale !?

Autres critères :

- Hydramios
- Anasarque
- Déviation médiastinale
- Compression




*Delacourt 2019*

# *Critères pronostics et lieu de naissance*

- Discussion Collégiale du lieu de naissance
- Pas de césarienne de principe
- Naissance à terme

**The Utility of the Congenital Pulmonary Airway Malformation-Volume Ratio in the Assessment of Fetal Echogenic Lung Lesions: A Systematic Review**

Stefan C. Kane[a–c] Emanuele Ancona[a, b, d] Karen L. Reidy[a, b] Ricardo Palma-Dias[a–c]

**Prenatal natural history of congenital pulmonary malformations: MALFPULM population-based cohort study**

*Ultrasound Obstet Gynecol* 2019; 54: 381–388

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**Des situations « Simples » :**

Compressions médiastinales
Epanchements et dysfonction cardiaque fœtale

**Dans les autres cas : CVR**

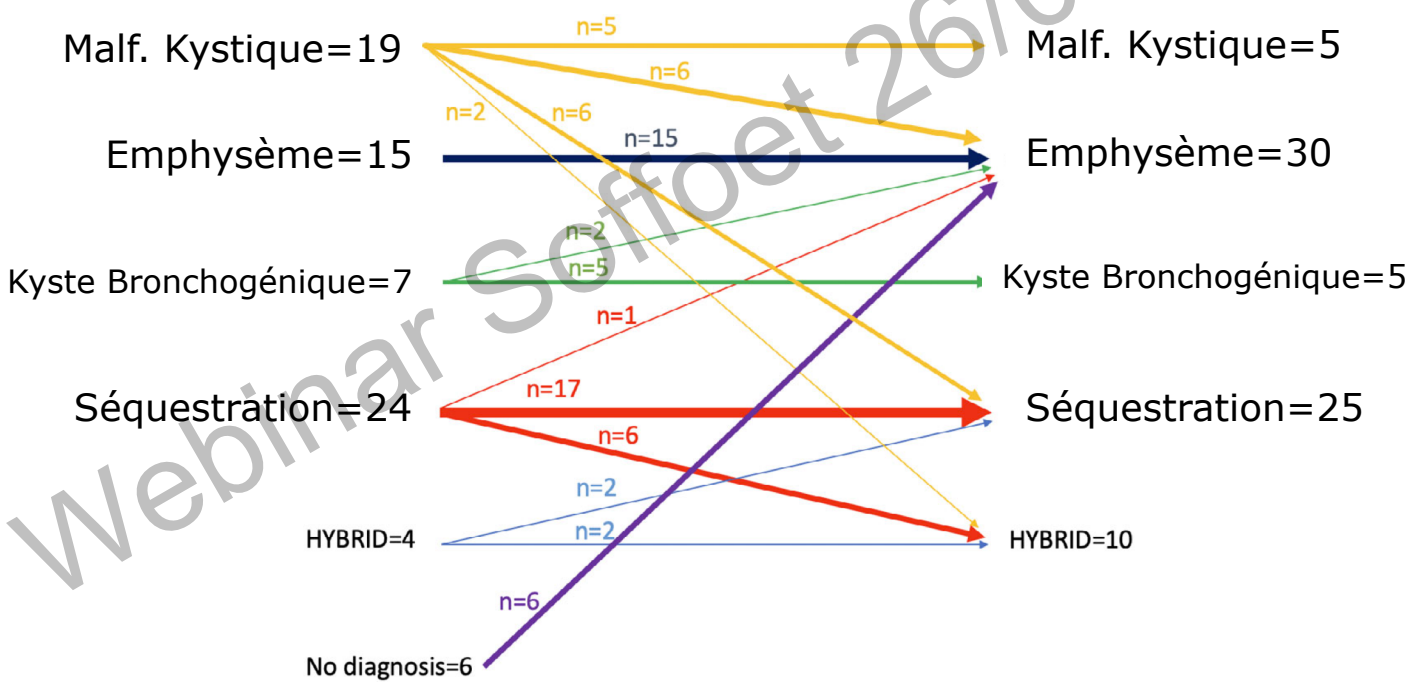
MALFPULM : Seuil = 0,4
Modification suivant le type lésionnel ??
0,9 ➜ 1,6

Ultrasound detected prenatal hyperechoic lung lesions and concordance with postnatal findings: A common aspect for multiple diagnoses
*Prenatal Diagnosis*. 2021;41:323–331.

Michela Cing Yu Wong[1] | Valentine Faure Bardon[2,3] | Konstantinos Farmakis[1] | Laureline Berteloot[4] | Alexandre Lapillonne[3,5] | Christophe Delacourt[6] | Sabine Sarnacki[1,3] | Yves Ville[2,3] | Naziha Khen-Dunlop[1,3]

# *Performance Diagnostique Anténatale*



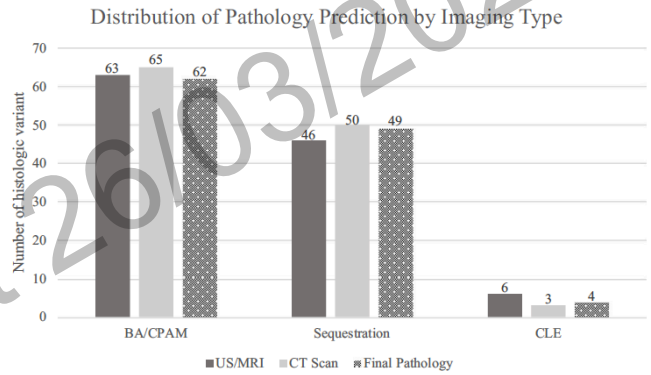


# Performance Diagnostique Anténatale

Accuracy of prenatal and postnatal imaging for management of congenital lung malformations

Journal of Pediatric Surgery 55 (2020) 844–847

Candace C. Style, MD [a,b], Amy R. Mehollin-Ray [a,c], Mariatu A Verla, MD [a,b], Oluyinka O. Olutoye, MD, PhD [a,b,d], Patricio E. Lau, MD [b], Brittany L. Johnson, MD [a,b], Alice King, MD [a,b], Sundeep G. Keswani, MD [a,b], Timothy C. Lee, MD [a,b,*]






Diagnostic accuracy of prenatal US, fetal MRI, and postnatal CT in infants with CLMs.

| | | Diagnostic accuracy | Sensitivity | Specificity | PPV | NPV |
|---|---|---|---|---|---|---|
| US/Fetal MRI* | Lesion Location (Left vs Right) | 100% | 100% | 100% | 100% | 100% |
| | Presence of Systemic Vessel | 89% | 88% | 90% | 87% | 90% |
| | Bronchial Atresia & CPAMs | 88% | 94% | 82% | 83% | 94% |
| | Bronchopulmonary Sequestration | 90% | 87% | 92% | 89% | 91% |
| | Congenital lobar emphysema | 97% | 75% | 98% | 60% | 99% |
| Postnatal CT | Lesion Location (Left vs Right) | 100% | 100% | 100% | 100% | 100% |
| | Presence of Systemic Vessel | 89% | 88% | 91% | 91% | 85% |
| | Bronchial Atresia & CPAMs | 94% | 96% | 92% | 92% | 96% |
| | Bronchopulmonary Sequestration | 87% | 90% | 88% | 90% | 88% |
| | Congenital lobar emphysema | 99% | 75% | 100% | 100% | 99% |

* US/MRI were performed and interpreted as paired imaging; MRI=magnetic resonance imaging; CT=computed topography; BPS=bronchopulmonary sequestration (intra- and extralobar), and CLE=congenital lobar emphysema; PPV: positive predictive value; NPV: negative predictive value.