Hernie de coupole diaphragmatique.

Que retenir depuis Mai 2018?

A.Benachi, J. Boubnova, L.Storme

Centre de Référence Maladie Rare: Hernie de Coupole Diaphragmatique

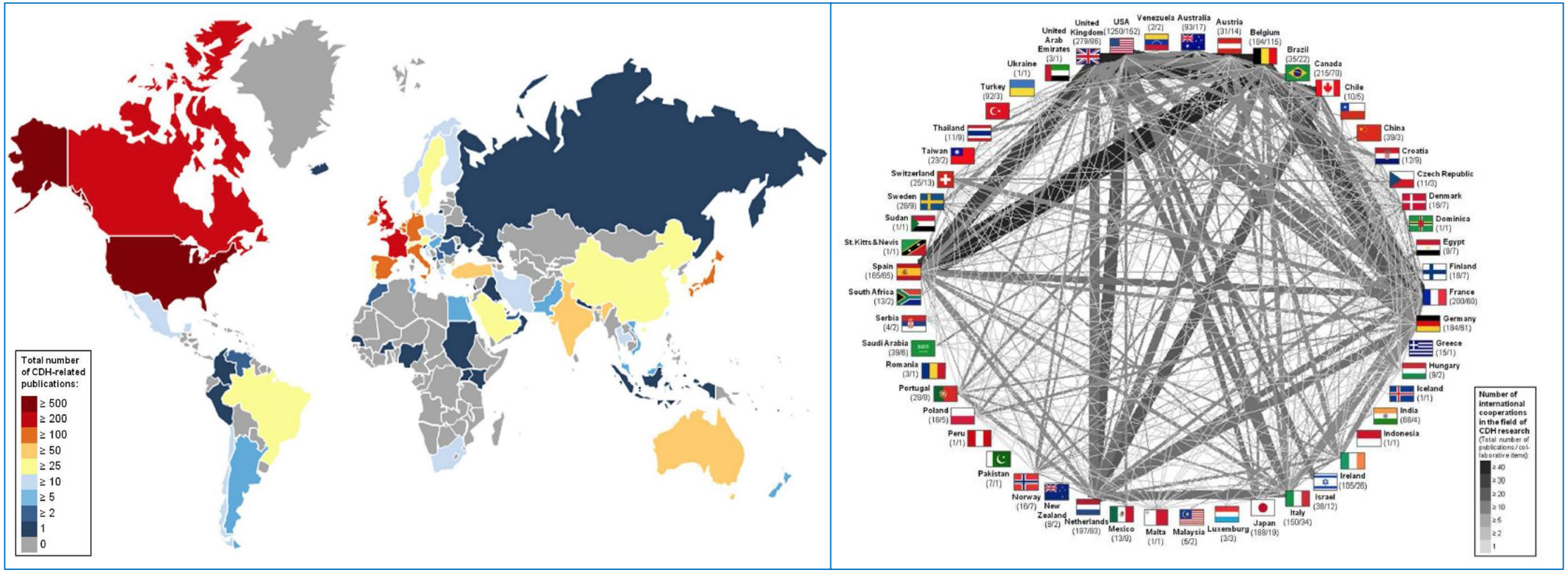
filière de santé maladies rares

FIMATHO
Filière des maladies rares abdomino-thoraciques

CDH Euro-Consortium

European Reference Network
for rare or low prevalence complex diseases
Network Inherited and Congenital Anomalies (ERNICA)

- 314 papiers
  - Prénatal, génétique, recherche fondamentale (66)
  - Techniques chirurgicales et anesthésiques (28)
  - Réanimation (60)
  - Suivi à long terme (26)
  - Parents – Qualités de vie (7)



- 3669 publications on CDH were identified, originating from 76 countries
- Globally, the largest number of scientific articles relating to CDH was published by the USA [n = 1250; (34.1%)], the UK [n = 279; (7.6%)], and Canada [n = 215; (5.9%)]. Most CDH papers were written in English [n = 3432; (93.5%)], followed by French [n = 87; (2.4%)] and German[n = 81; (2.2%)]

Ped Surg Int (2018) 34:907-917

Table 1 Ten most productive national and international CDH cooperations and registries

Rank	Name of cooperation or registry	Year of foundation	Participating centers	Extent of collaboration	Publications
1	CDH Study Group (CDHSG)	1995	$n=112$	14 countries worldwide	$n=35$
2	Canadian Pediatric Surgery Network (CAPSNet)	2005	$n=17$	Canada nationwide	$n=14$
3	FETO Task Group	2004	$n=5$	4 European countries, 1 U.S. center	$n=6$
4	CDH EURO Consortium	2008	$n=22$	13 European countries, 1 Canadian center	$n=5$
5	French CDH Study Group/Center for Rare Diseases—CDH	2007	$n=31$	France nationwide	$n=5$
6	Japanese CDH Study Group	2011	$n=9$	Japan nationwide	$n=5$
7	Antenatal CDH Registry Group	2005	$n=10$	5 European countries, 1 U.S. & 1 Israeli center	$n=3$
8	Groupe Radiopédiatrique de Recherche en Imagerie foetale (GRRIF)	1980	$n=10$	2 European countries	$n=2$
9	National Birth Defects Prevention Study (NBDPS)	1996	NS	10 U.S. states	$n=2$
10	Western Canadian ECMO Follow-Up Group	1997	$n=5$	Western Canada	$n=2$

Ped Surg Int (2018) 34:907-917

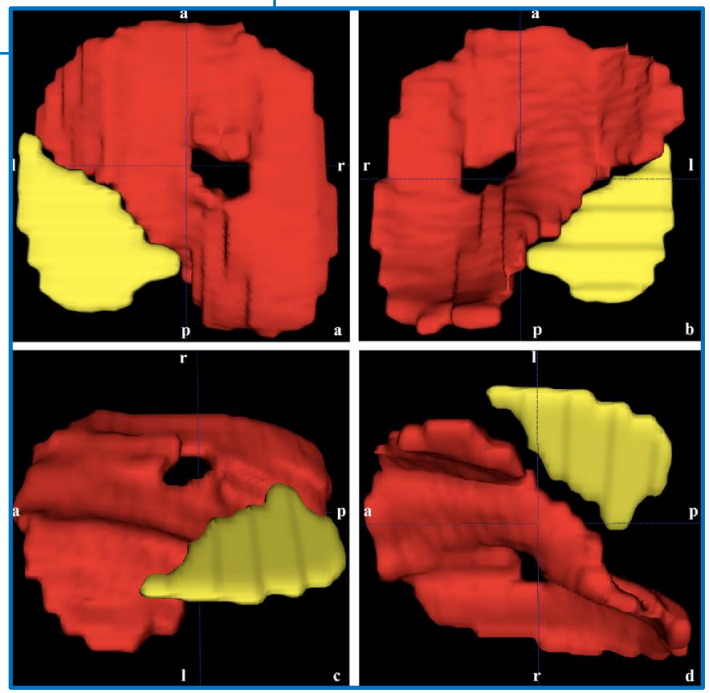
Prénatal et Génétique

3D reconstruction of diaphragmatic defects in CDH: a fetal MRI study

Short Title: Fetal MRI of CDH defects

Florian Prayer[1], Martin Metzelder[2], Wilfried Krois[2], Peter C. Brugger[3], Gerlinde M. Gruber[3], Michael Weber[1], Anke Scharrer[4], Alexander Rokitansky[5], Georg Langs[6], Daniela Prayer[1], Ewald Unger[7] and Gregor Kasprian[1]

- To assess the clinical feasibility and validity of fetal MRI-based 3D reconstructions to localize, classify, and quantify diaphragmatic defects in congenital diaphragmatic hernia
- Areas of the intact diaphragm and the defect were measured and defect-to-diaphragmatic ratios (DDR) were calculated
- The need for prosthetic patch repair and diaphragm growth dynamics, in cases with repeated in vivo fetal MRI scans, were analyzed based on DDR.



Ultrasound Obstet Gynecol (2019) April

Prénatal et Génétique

Proposal for standardized prenatal ultrasound assessment of the fetus with congenital diaphragmatic hernia by the European reference network on rare inherited and congenital anomalies (ERNICA)

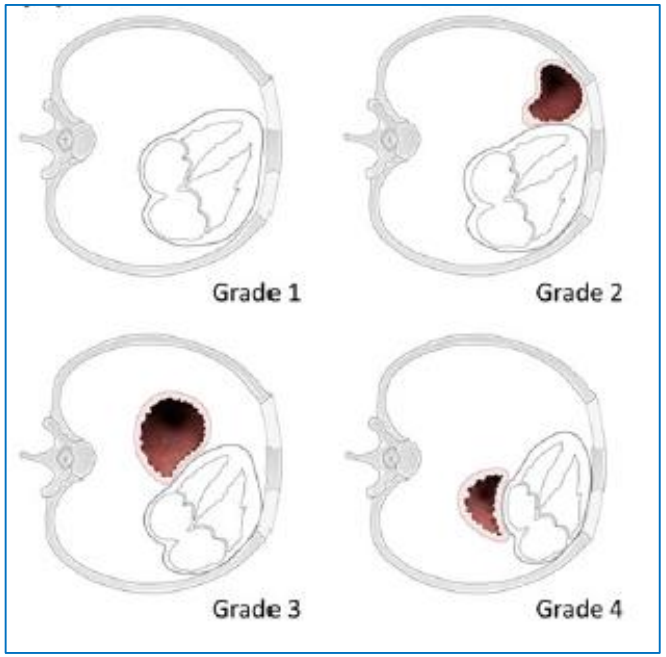
Francesca Maria Russo[1,2] | Anne-Gael Cordier[3] | Luc De Catte[1,2] | Julien Saada[4] | Alexandra Benachi[3,4] | Jan Deprest[1,2,5] |
on behalf of the Workstream Prenatal Management, ERNICA European reference network

What's already known about this topic?

- Congenital diaphragmatic hernia is associated with high postnatal mortality and morbidity.
- Prenatal diagnosis is often possible with ultrasound, after which patients are referred to specialized centers.
- In isolated cases, antenatal ultrasound allows prediction of outcome.

What does this study add?

- We provide a practical and instructional guide for the standardized assessment of fetuses with isolated left or right congenital diaphragmatic hernia and individualized prediction of neonatal outcome.



Prenat Diagn 2018;38:629-637

Prénatal et Génétique

Contents lists available at ScienceDirect

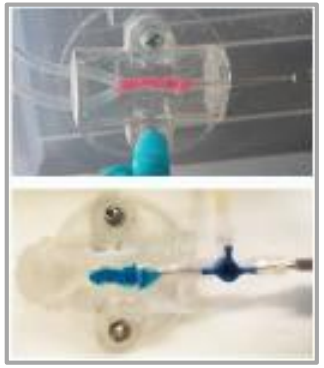
Materials Science & Engineering C

journal homepage: www.elsevier.com/locate/msec

An injectable, degradable hydrogel plug for tracheal occlusion in congenital diaphragmatic hernia (CDH)

Chiara Emma Campiglio[a,d], Martina Villonio[a], Raffaele L. Dellacà[b], Fabio Mosca[c], Lorenza Draghi[a,d,*]

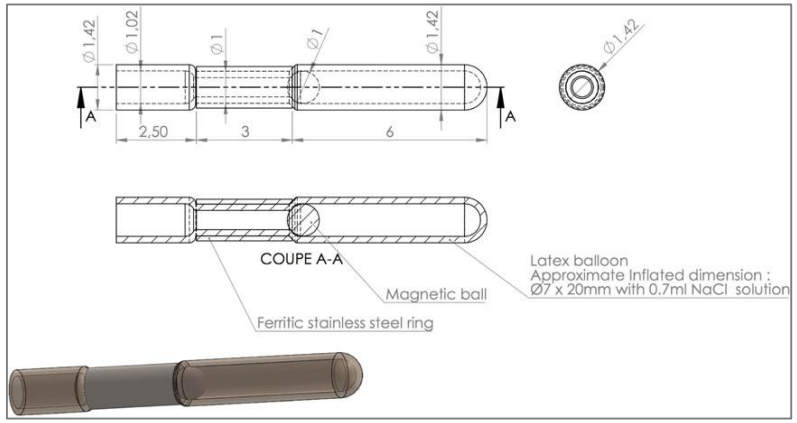
Mat Science Eng 2019, 99, 430-439



Evaluation of a new balloon for fetal endoscopic tracheal occlusion in the nonhuman primate model

Nicolas Sananès[1,2] | Pierrick Regnard[3] | Nicolas Mottet[1] | Claire Miry[1] | Lyne Fellmann[3] | Laure Haelewyn[3] | Maïa Delaine[1] | Anne Schneider[4] | Christian Debry[2,5] | Romain Favre[1]

Prenat Diagn 2019,

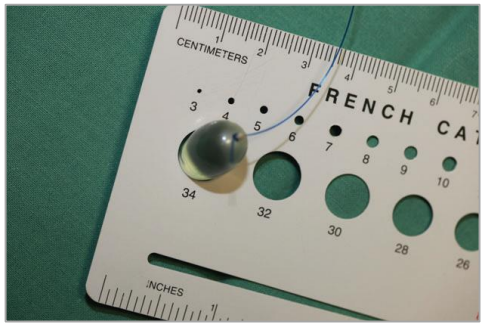


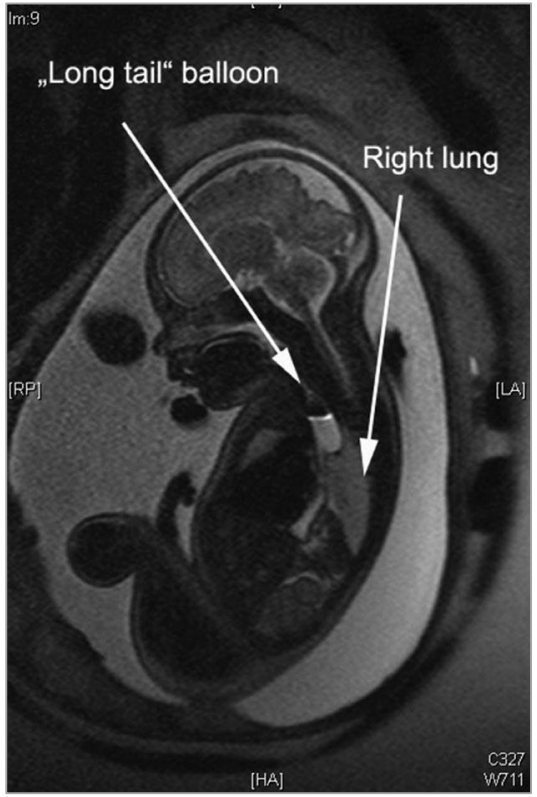
Prénatal et Génétique

Long tail balloon as a new approach for fetoscopic tracheal occlusion for a treatment of severe congenital diaphragmatic hernia

Michael Tchirikov[1], Carsten Springer[2], Sven Seeger[3], Curd Behrmann[4], Michael Bergner[1] and Roland Haase[5]

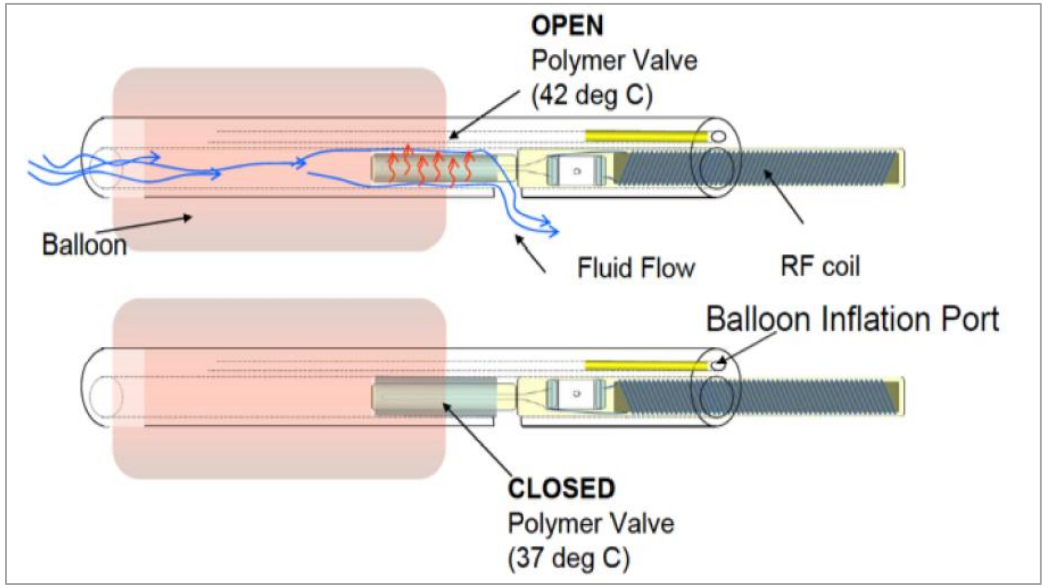
J Obstet Gyneacol Res 2019, 45:719-723





Characterization of a reversible thermally-actuated polymer-valve: A potential dynamic treatment for congenital diaphragmatic hernia

Justin S. Baba[1,2]*, Timothy E. McKnight[1], M. Nance Ericson[1], Anthony Johnson[3¤], Kenneth J. Moise Jr.[3¤], Boyd M. Evans III[1]



Plos One 2018, Dec 27

Prénatal et Génétique

Sildenafil for Antenatal Treatment of Congenital Diaphragmatic Hernia: From Bench to Bedside

Francesca M. Russo[1], Felix De Bie[1], Ryan Hodges[2], Alan Flake[3] and Jan Deprest[1,4,*]

Conclusion: There is preclinical evidence that maternally administered sildenafil prevents the vascular changes that lead to PPH in CDH newborns. The phase I/IIb clinical study together with the pregnancy-Physiologically Based PharmacoKinetic model will define the maternal dose needed for a therapeutic effect in the foetus. Foetal safety will be investigated both in the clinical study and in the sheep. The final step will be a multicentre, randomized, placebo-controlled trial.

Current Pharmaceutical Design. 2019; 25, 1-8

Lactation Experience of Mothers and Feeding Outcomes of Infants with Congenital Diaphragmatic Hernia

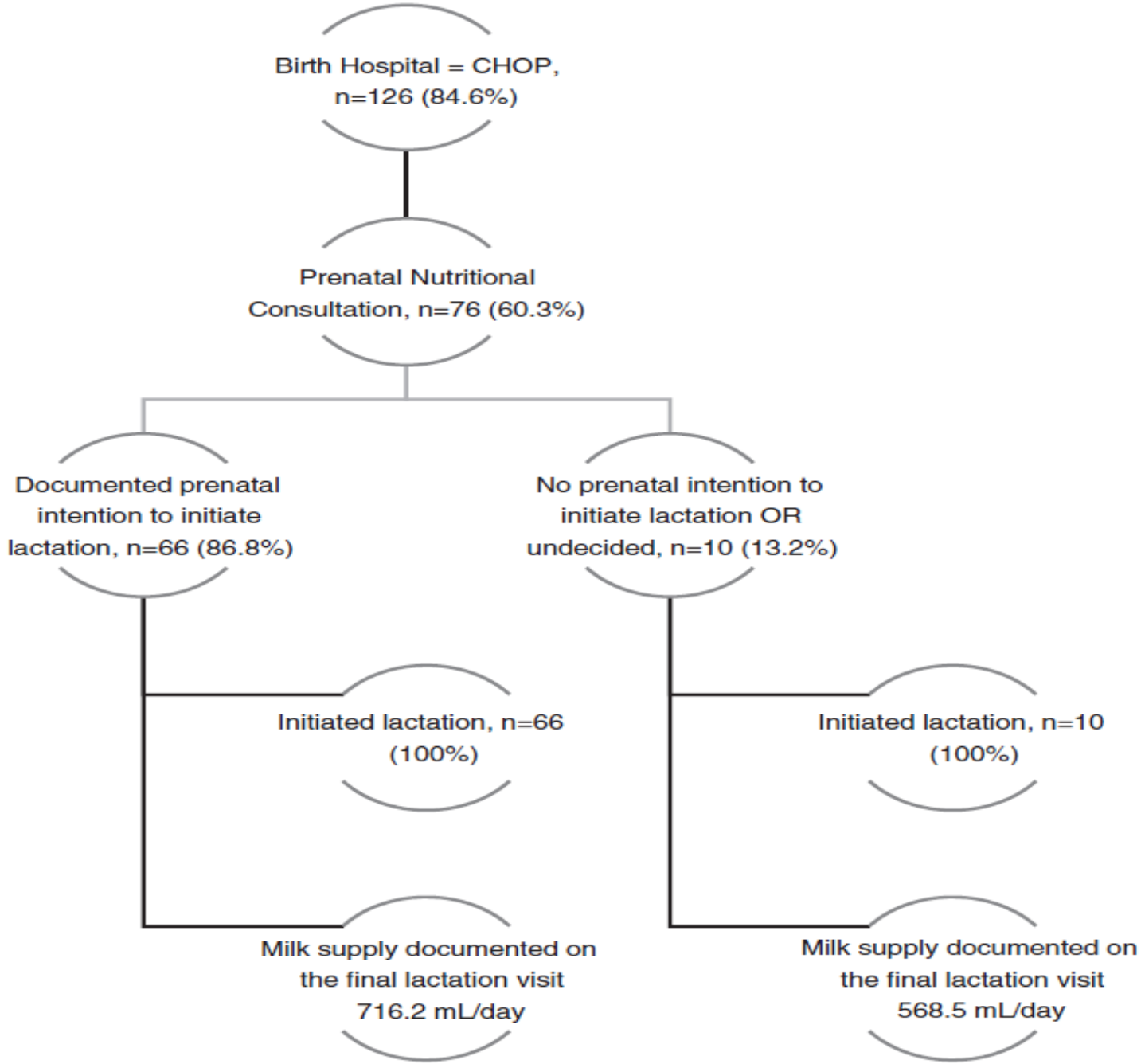
Diane L. Spatz,[1,2] Elizabeth B. Froh,[1,2] Dana Bartholomew,[3] Taryn Edwards,[4] Katherine T. Wild,[5] Holly Hedrick,[4] and Ursula Nawab[5]

TABLE 1. DEMOGRAPHICS

	n	*Mean ± SD*	*Median (IQR)*	*Range*
Gestational age, weeks	149	37.73 ± 1.73	38 (37–39)	31–41
Birth weight, kg	149	3.13 ± 0.51	3.1 (2.8–3.5)	1.5–4.4
Day of life of admission, days	149	2.03 ± 6.21	1 (1–1)	1–63
Total parental nutrition, days	149	33.74 ± 25.52	28 (16–43)	4–161
Duration of ventilation, days	149	26.99 ± 26.18	21 (9–35)	0–202
Length of stay, days	149	69.18 ± 58.51	58 (28–86)	6–343

IQR, interquartile range; SD, standard deviation.

Breastfeeding Medicine 2019

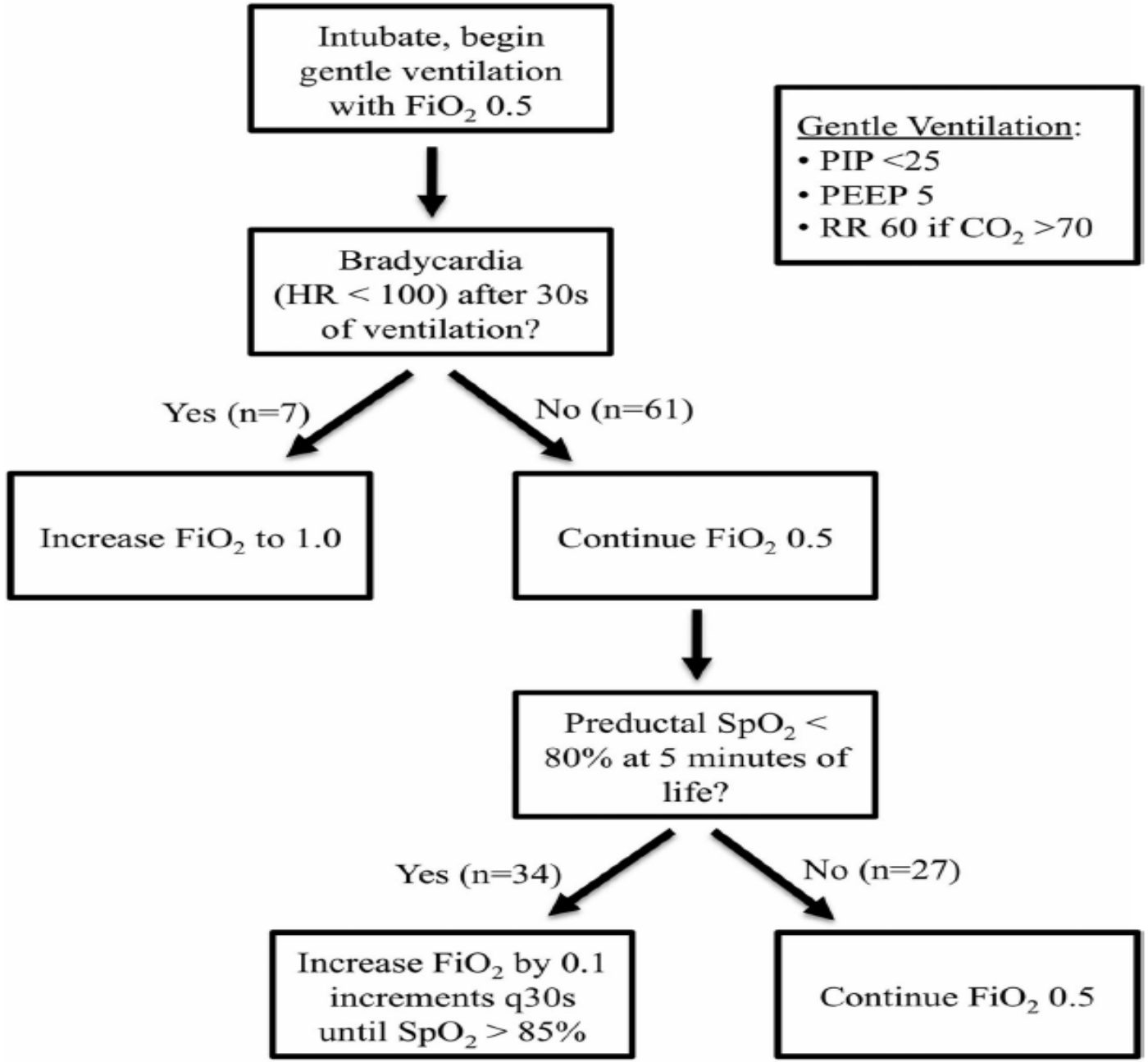

Birth Hospital = CHOP, n=126 (84.6%)
Prenatal Nutritional Consultation, n=76 (60.3%)
Documented prenatal intention to initiate lactation, n=66 (86.8%)
No prenatal intention to initiate lactation OR undecided, n=10 (13.2%)
Initiated lactation, n=66 (100%)
Initiated lactation, n=10 (100%)
Milk supply documented on the final lactation visit 716.2 mL/day
Milk supply documented on the final lactation visit 568.5 mL/day

Journal of Perinatology
https://doi.org/10.1038/s41372-017-0031-5

ARTICLE

Reduced oxygen concentration for the resuscitation of infants with congenital diaphragmatic hernia

John S. Riley[1] · Ryan M. Antiel[1] · Natalie E. Rintoul[1] · Anne M. Ades[1] · Lindsay N. Waqar[1] · Nan Lin[2] · Lisa M. Herkert[1] · Jo Ann D'Agostino[3] · Casey Hoffman[3] · William H. Peranteau[1] · Alan W. Flake[1] · N. Scott Adzick[1] · Holly L. Hedrick[1]


Intubate, begin gentle ventilation with FiO_2 0.5
Gentle Ventilation:
• PIP <25
• PEEP 5
• RR 60 if CO_2 >70
Bradycardia (HR < 100) after 30s of ventilation?
Yes (n=7)
No (n=61)
Increase FiO_2 to 1.0
Continue FiO_2 0.5
Preductal SpO_2 < 80% at 5 minutes of life?
Yes (n=34)
No (n=27)
Increase FiO_2 by 0.1 increments q30s until SpO_2 > 85%
Continue FiO_2 0.5

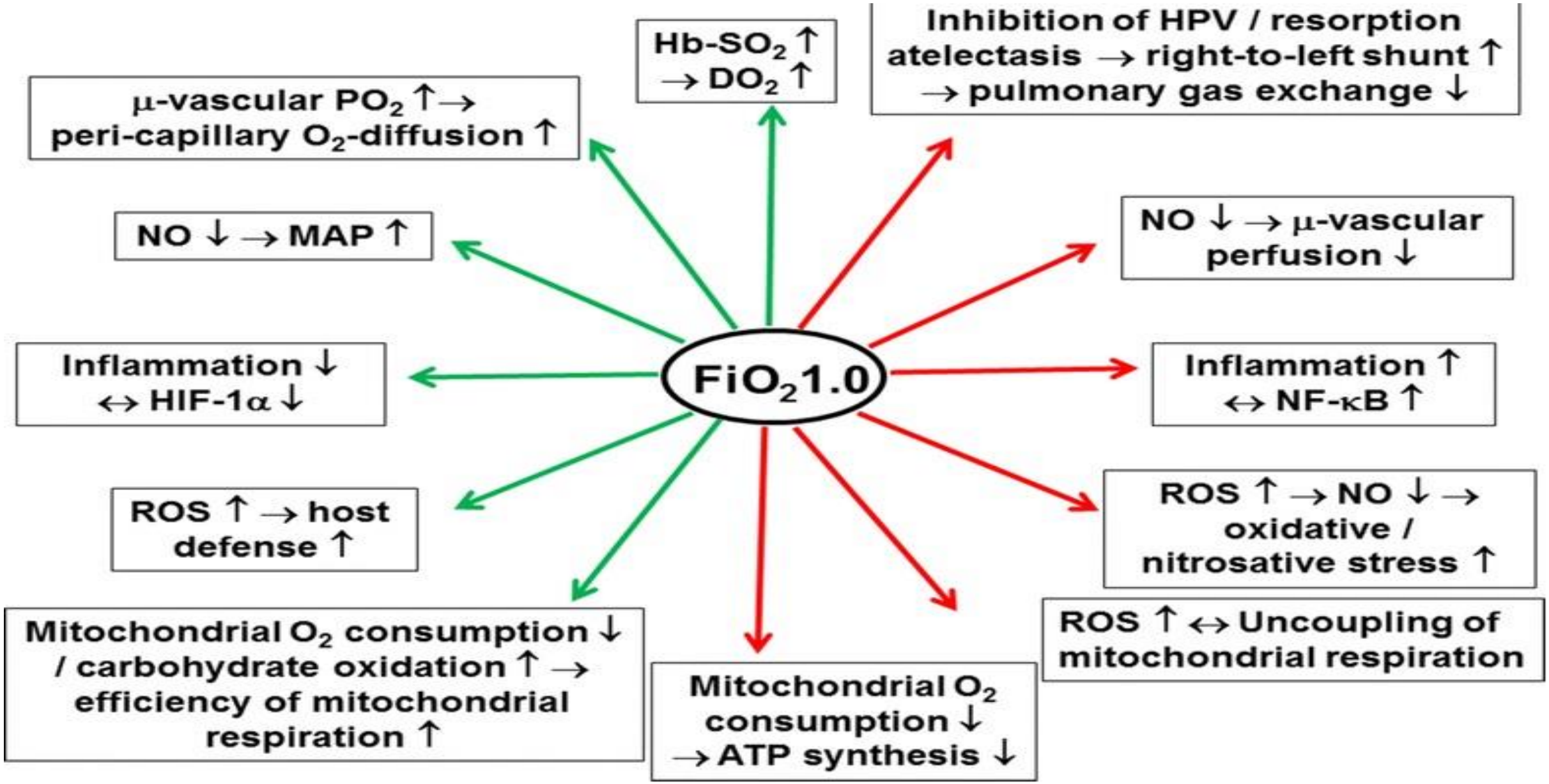
Variable	Historical controls $n = 45$	Study cohort $n = 68$	P value
Bradycardia	2 (4.4)	7 (10.3)	0.313
Chest compressions	0 (0)	1 (1.5)	0.598
Epinephrine	0 (0)	0 (0)	1
MAP ever < 40 mmHg	7 (15.6)	12 (17.6)	0.771
Fluid bolus	13 (28.9)	26 (38.2)	0.306
Dopamine	1 (2.2)	0 (0)	0.398
Apgar—1 min[a]	4.5 (3)	5 (5)	0.648
Apgar—5 min[a]	7 (2)	8 (3)	0.193
Maximum FiO_2			
Resuscitation	1.0 (0)	0.75 (0.24)	**<0.001**
Day of life 1[c]	0.85 (0.22)	0.58 (0.22)	**<0.001**
Day of life 2[c]	0.66 (0.28)	0.48 (0.23)	**0.0043**
Day of life 3[c]	0.48 (0.21)	0.48 (0.21)	0.985
Lactate			
Day of life 1	2.66 (2.35)	2.25 (1.21)	0.36
Day of life 2	2.01 (1.51)	1.69 (0.66)	0.273
Day of life 3	1.72 (1.23)	1.39 (0.46)	0.171

Devenir

Table 3 Univariate analysis of primary outcomes

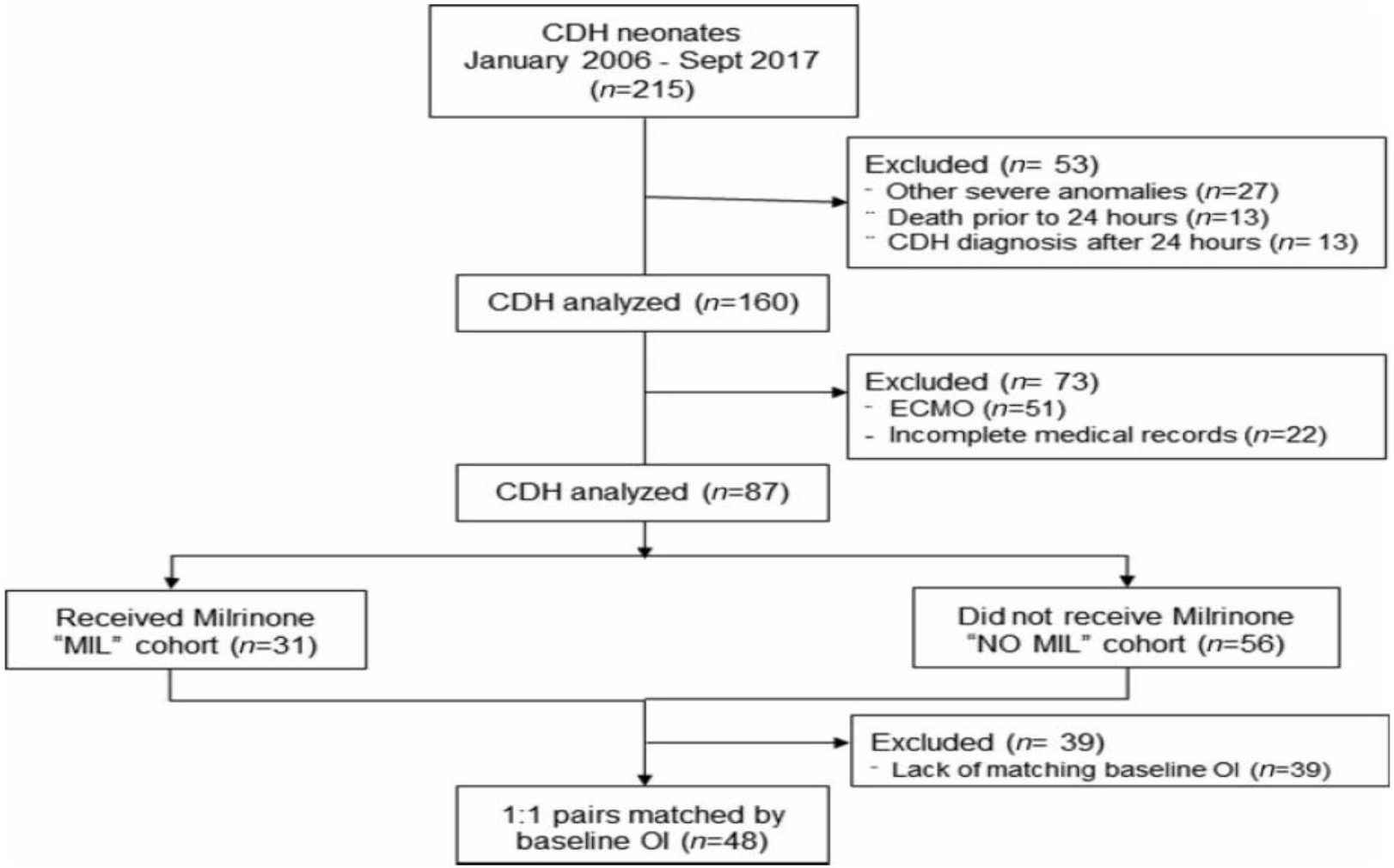
Variable	Historical controls $n = 45$	Study cohort $n = 68$	P value
Survival	33 (73.3)	57 (83.8)	0.175
Days intubated	28.3 (19.2)	39.0 (41.5)	0.0796
ECMO initiated	19 (42.2)	17 (25.0)	0.0540
Days on ECMO	17.2 (7.4)	19.8 (11.9)	0.446
Days to surgery	16.3 (14.1)	18.2 (15.0)	0.538

Reiley, J Perinat 2018


Hb-SO2 ↑ → DO2 ↑
Inhibition of HPV / resorption atelectasis → right-to-left shunt ↑ → pulmonary gas exchange ↓
μ-vascular PO2 ↑ → peri-capillary O2-diffusion ↑
NO ↓ → μ-vascular perfusion ↓
NO ↓ → MAP ↑
FiO2 1.0
Inflammation ↓ ↔ HIF-1α ↓
Inflammation ↑ ↔ NF-κB ↑
ROS ↑ → host defense ↑
ROS ↑ → NO ↓ → oxidative / nitrosative stress ↑
ROS ↑ ↔ Uncoupling of mitochondrial respiration
Mitochondrial O2 consumption ↓ / carbohydrate oxidation ↑ → efficiency of mitochondrial respiration ↑
Mitochondrial O2 consumption ↓ → ATP synthesis ↓

Is Milrinone Effective for Infants with Mild-to-Moderate Congenital Diaphragmatic Hernia?

Michelle Mears, DNP[1,2] Michelle Yang, MD[2,3] Bradley A. Yoder, MD[2,3]

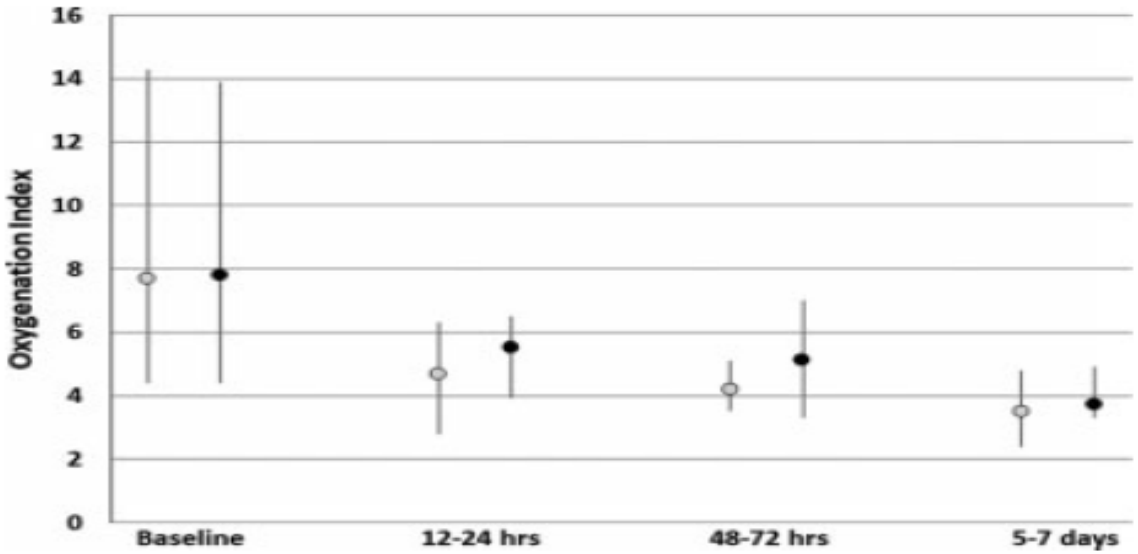


Am J Perinatology 2019

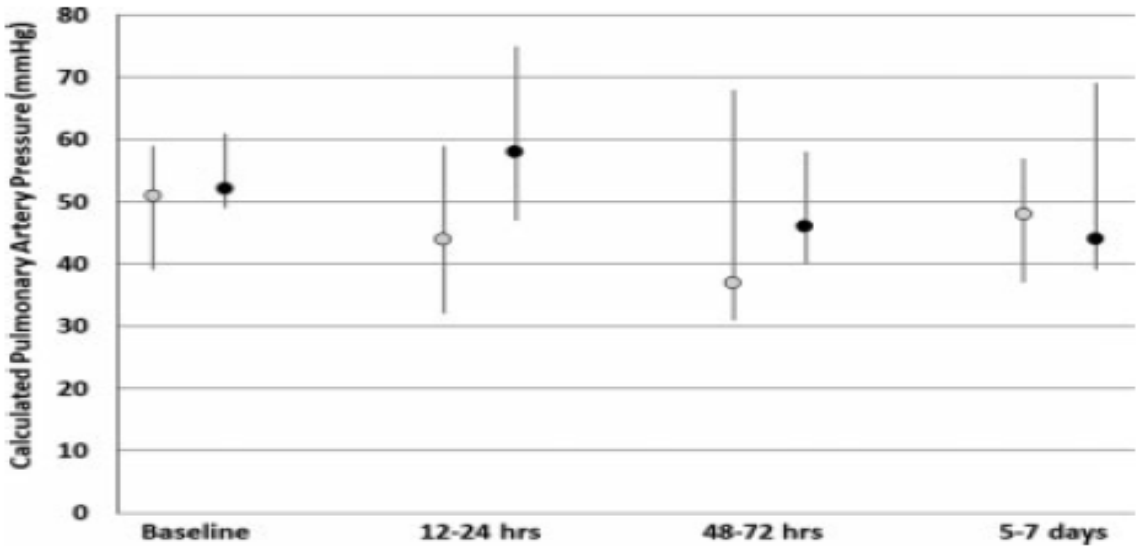
Table 1 Demographic features of milrinone treated versus nontreated CDH neonates

	No milrinone therapy ($n = 24$)	Milrinone treated ($n = 24$)	*p*-Value
Gestational age, weeks	38.2 ± 1.5	38.1 ± 1.5	0.324
Birth weight, grams	3,134 ± 456	3,064 ± 409	0.580
Male	16 (67%)	15 (63%)	0.763
Fetal diagnosis	13 (54%)	18 (75%)	0.227
Age at repair, hours	68 (30)	92 (49)	0.048
Left-sided	22 (92%)	21 (88%)	1.00
Defect size > 2	7 (29%)	7 (29%)	1.00
Patch repair	2 (8%)	5 (21%)	0.193
Baseline OI	9.6 + 6.7	9.5 ± 6.5	0.964
Baseline PAP, mm Hg	49 ± 11	53 ± 11	0.327
Bidirectional or right-to-left across initial PDA	15 (63%)	20 (83%)	0.193
Inhaled NO	19 (79%)	19 (79%)	1.00
Dopamine	16 (67%)	22 (92%)	0.072
Hydrocortisone	19 (79%)	18 (75%)	0.712

Index d’Oxygénation

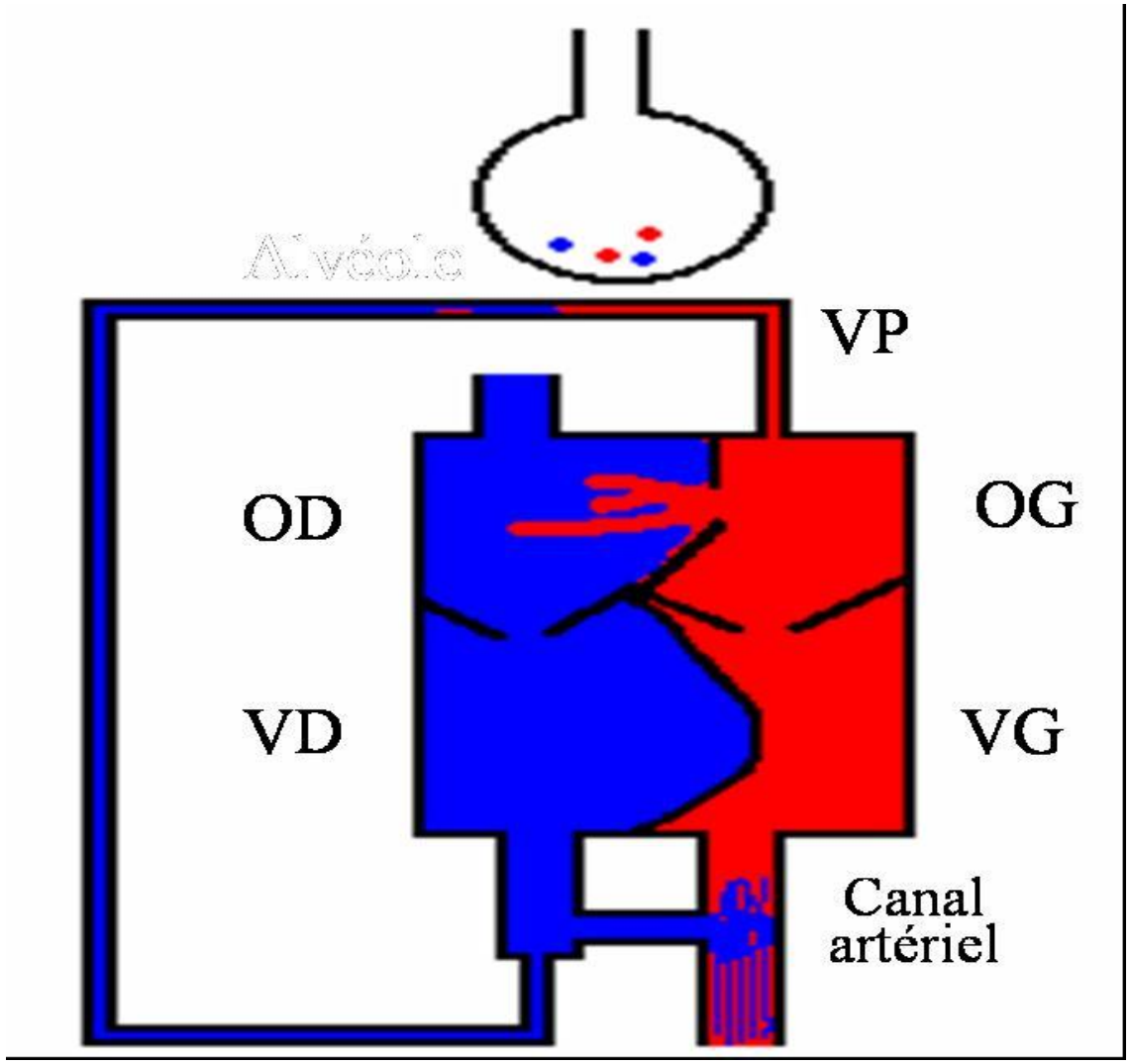


Pression artérielle pulmonaire

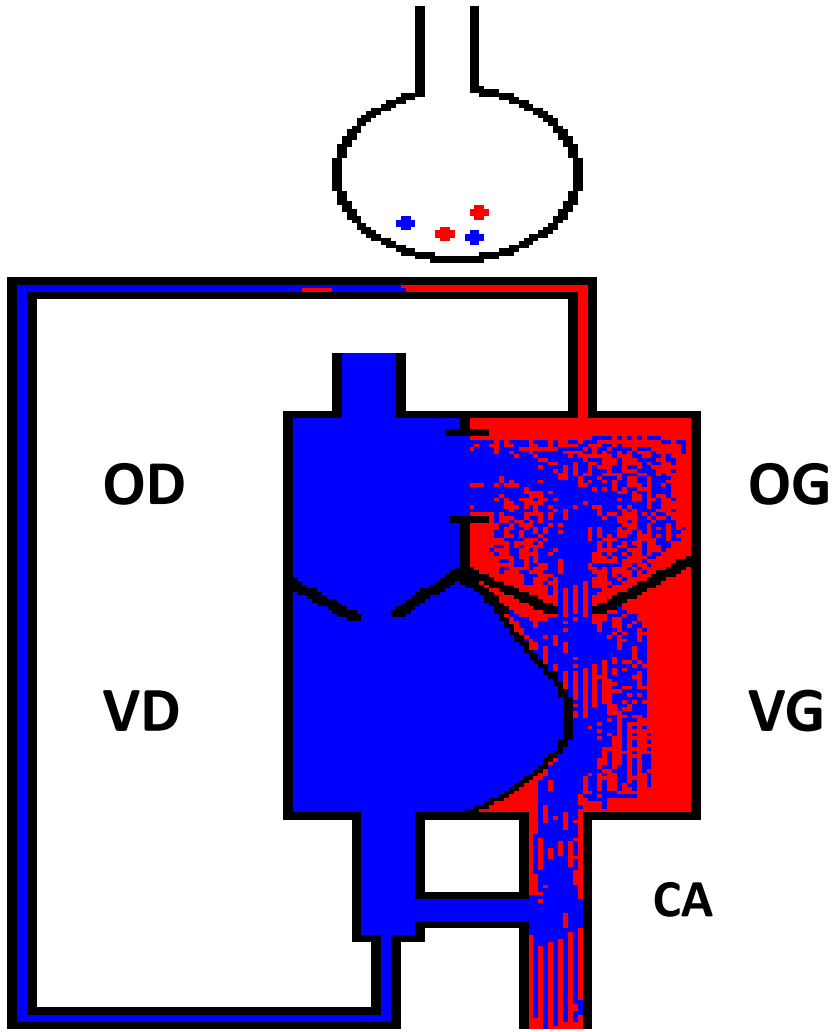


$PAP = (Q_{pulm} \times PVR) + LAP$

1. PPHN + L to R shunting FO



2. PPHN + R to L shunting FO



FIMATHO
Filière des maladies rares abdomino-thoraciques

Hernie de coupole diaphragmatique
Centre de Maladie Rare

Top 3 publications chirurgie

Mai 2018 - Mai 2019

Dr Julia Boubnova

Chirurgie pédiatrique

CHU Timone Enfants

Marseille

Journée nationale HDC, Paris le 16 mai 2019

European Reference Networks

ORIGINAL ARTICLE

Preoperative sonographic evaluation of the defect size and the diaphragm rim in congenital diaphragmatic hernia — preliminary experience

Kengo Hattori[1] · Shigeru Takamizawa[1] · Yuichiro Miyake[1] · Tomoko Hatata[1] · Katsumi Yoshizawa[1] · Tomoko Furukawa[2] · Yoshiaki Kondo[2]

Operative approach	Defect size (mm)		Herniated organs	
	Ultrasound	intraoperative	Ultrasound	Intraoperative
Thoracoscopic	23 × 25	20 × 26	Bowel	Bowel
Thoracoscopic	23 × 30	20 × 30	Bowel	Bowel
Thoracoscopic	25 × 43	30 × 30	Bowel, spleen	Bowel, spleen
Thoracoscopic	21 × 23	20 × 25	Bowel	Bowel
Thoracoscopic	19 × 24	10 × 30	Bowel	Bowel
Open	32 × 33	30 × 50	Bowel, spleen	Bowel, spleen
Open	Total absence	40 × 50	Bowel, stomach, spleen	Bowel, stomach, spleen

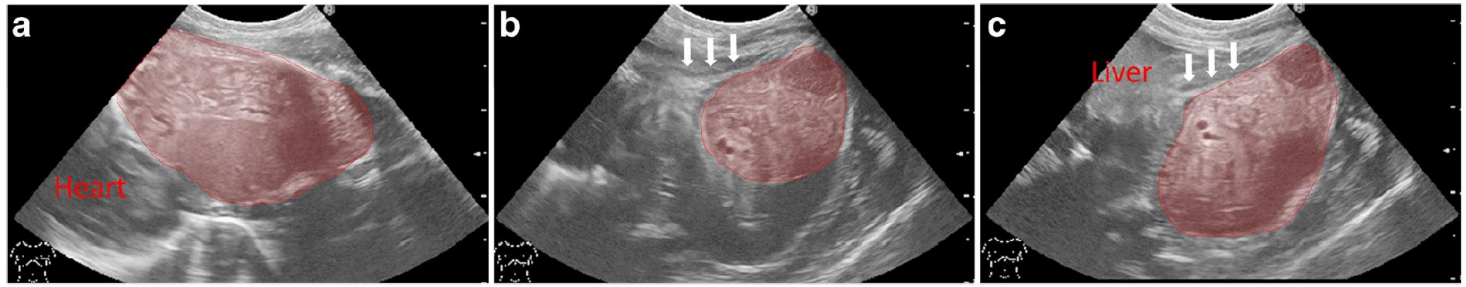


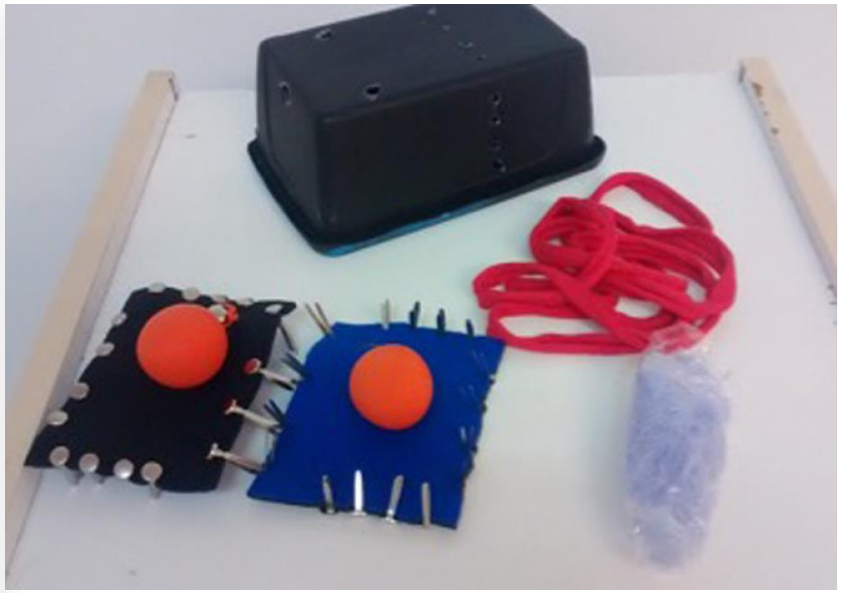
Fig. 3 Preoperative ultrasound images of the herniated abdominal contents in the axial plane. **a** Image at the level of the heart demonstrates intrathoracic herniated contents (*red shaded area*). **b** Image at the narrowest point shows the diaphragmatic defect (*red shaded area*). **c** Image at the level of abdomen shows the fragment of the diaphragm (*arrows*) and the herniated structures (*red shaded area*)

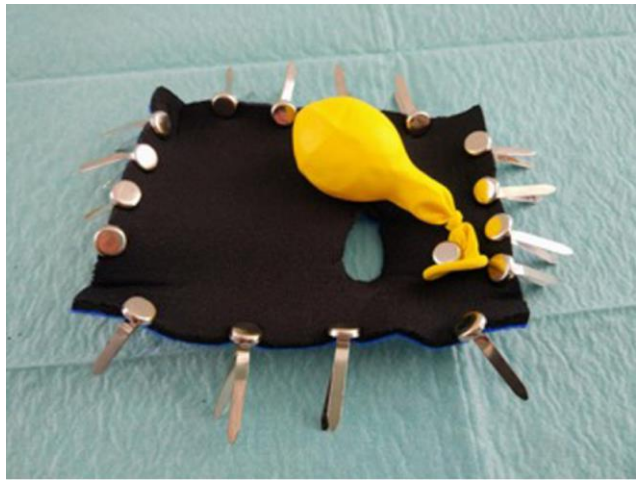
- Etude pilote pour la concordance de mesure de la taille du défect à l'écho et les constatations opératoires
- Bonne corrélation
- Echographiste +++

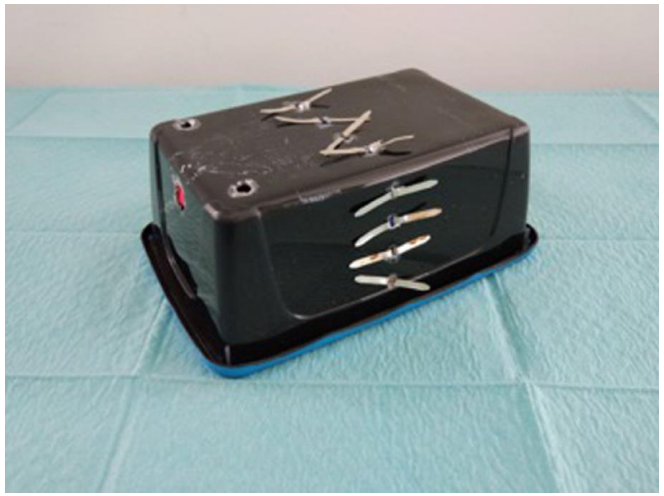
Validation of a Low-Cost Do-It-Yourself Model for Neonatal Thoracoscopic Congenital Diaphragmatic Hernia Repair

Pedro Reino-Pires, MD, MSc, and Manuel Lopez, PhD†*

*Pediatric Surgery Department, Hospital de Dona Estefânia, Lisbon, Portugal; and †Pediatric Surgery Department, Hospital Universitario de Vall d'Hebron, Universidad Autónoma de Barcelona, Barcelona, España







This study shows that this simple model is reproducible and realistic enough to simulate a neonatal thoracoscopic CDH repair. Additionally, it can be perceived as a platform prone to modification as one sees fit, having the potential to be used for other simulations. Therefore, with these characteristics and considering its low cost, easy built features, and access to materials, this model is a good tool for every pediatric surgeon and a general contribution to pediatric surgery simulation.

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Potential survival benefit with repair of congenital diaphragmatic hernia (CDH) after extracorporeal membrane oxygenation (ECMO) in select patients: Study by ELSO CDH Interest Group

Patrick T. Delaplain [a,b], Matthew T. Harting [c], Tim Jancelewicz [d], Lishi Zhang [e], Peter T. Yu [b,f], Matteo Di Nardo [g], Yanjun Chen [e], James E. Stein [a], Henri R. Ford [a,h], Danh V. Nguyen [i], Yigit Guner [b,f,*]

Purpose: Studying the timing of repair in CDH is prone to confounding factors, including variability in disease severity and management. We hypothesized that delaying repair until post-ECMO would confer a survival benefit.

Methods: Neonates who underwent CDH repair were identified within the ELSO Registry. Patients were then divided into on-ECMO versus post-ECMO repair. Patients were 1:1 matched for severity based on pre-ECMO covariates using the propensity score (PS) for the timing of repair. Outcomes examined included mortality and severe neurologic injury (SNI).

Results: After matching, 2,224 infants were included. On-ECMO repair was associated with greater than 3-fold higher odds of mortality (OR 3.41, 95% CI: 2.84–4.09, $p<0.01$). The odds of SNI was also higher for on-ECMO repair (OR 1.49, 95% CI: 1.13–1.96, $p<0.01$). A sensitivity analysis was performed by including the length of ECMO as an additional matching variable. On-ECMO repair was still associated with higher odds of mortality (OR 2.38, 95% CI: 1.96–2.89, $p<0.01$). Results for SNI were similar but were no longer statistically significant (OR 1.33, 95% CI: 0.99–1.79, $p=0.06$).

Conclusions: Of the infants who can be liberated from ECMO and undergo CDH repair, there is a potential survival benefit for delaying CDH repair until after decannulation.

In summary, there appears to be a potential survival benefit associated with delaying repair until after ECMO as opposed to repair on-ECMO. This is similar to the findings of other single institution and retrospective database studies. However, it would not be accurate to condemn all repair on-ECMO, and the correct timing becomes less clear in certain infants, especially those infants that cannot be successfully weaned who may have the severest form of the disease. The results of this study found that for those infants that can be successfully weaned from ECMO, repair should be delayed until after decannulation.

Contents lists available at ScienceDirect

Biomaterials

journal homepage: www.elsevier.com/locate/biomaterials

Regeneration of diaphragm with bio-3D cellular patch

Xiu-Ying Zhang [a, *], Yusuke Yanagi [a], Zijing Sheng [b], Kouji Nagata [a], Koichi Nakayama [c, **], Tomoaki Taguchi [a]

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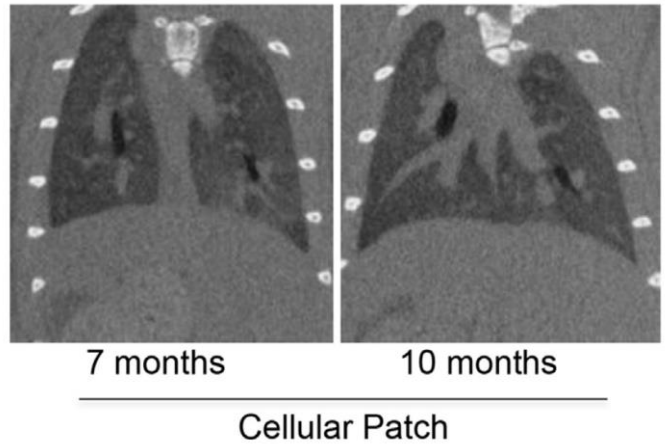
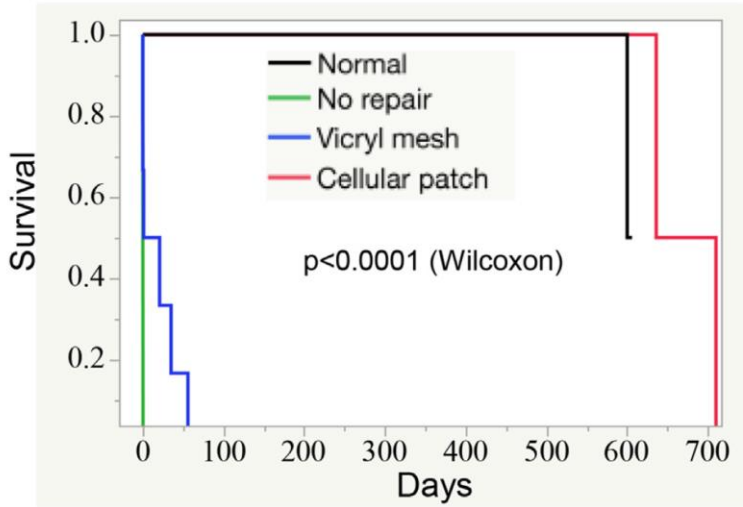


Neonates with congenital diaphragmatic hernia often require surgical defect closure with a patch. Alternatives to native diaphragmatic tissue are critically needed for this paediatric surgery. The clinical efficacy of mesh patches is limited by complications associated with residual foreign material and by hernia recurrence. In this study, we used a novel bio-3D printer method to generate large scaffold-free tissue patches composed of human cells. The resulting large tissue constructs had high elasticity and strength. Cellular patches were transplanted into rats with surgically created diaphragmatic defects. Rats survived for over 710 days after implantation of tissue constructs. CT confirmed complete tissue integration of the grafts during rat growth. Histology revealed regeneration of muscle structure, neovascularization, and neuronal networks within the reconstructed diaphragms. Our results demonstrate that created cellular patches are a highly safe and effective therapeutic strategy for repairing diaphragmatic defects, and thus pave the way for a clinical trial.



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