Hôpital Antoine-Béclère AP-HP

Hernie de coupole diaphragmatique

Grossesse et HCD

A.Benachi, L.Montalva

Centre Maladie Rare: Hernie de Coupole Diaphragmatique

Centre Pluridisciplinaire de Diagnostic Prénatal - Paris Saclay. France

Service de Chirurgie Pédiatrique - Hôpital Robert Debré. France

Inserm

CDH Euro-Consortium

FIMATHO

Filière des maladies rares abdomino-thoraciques

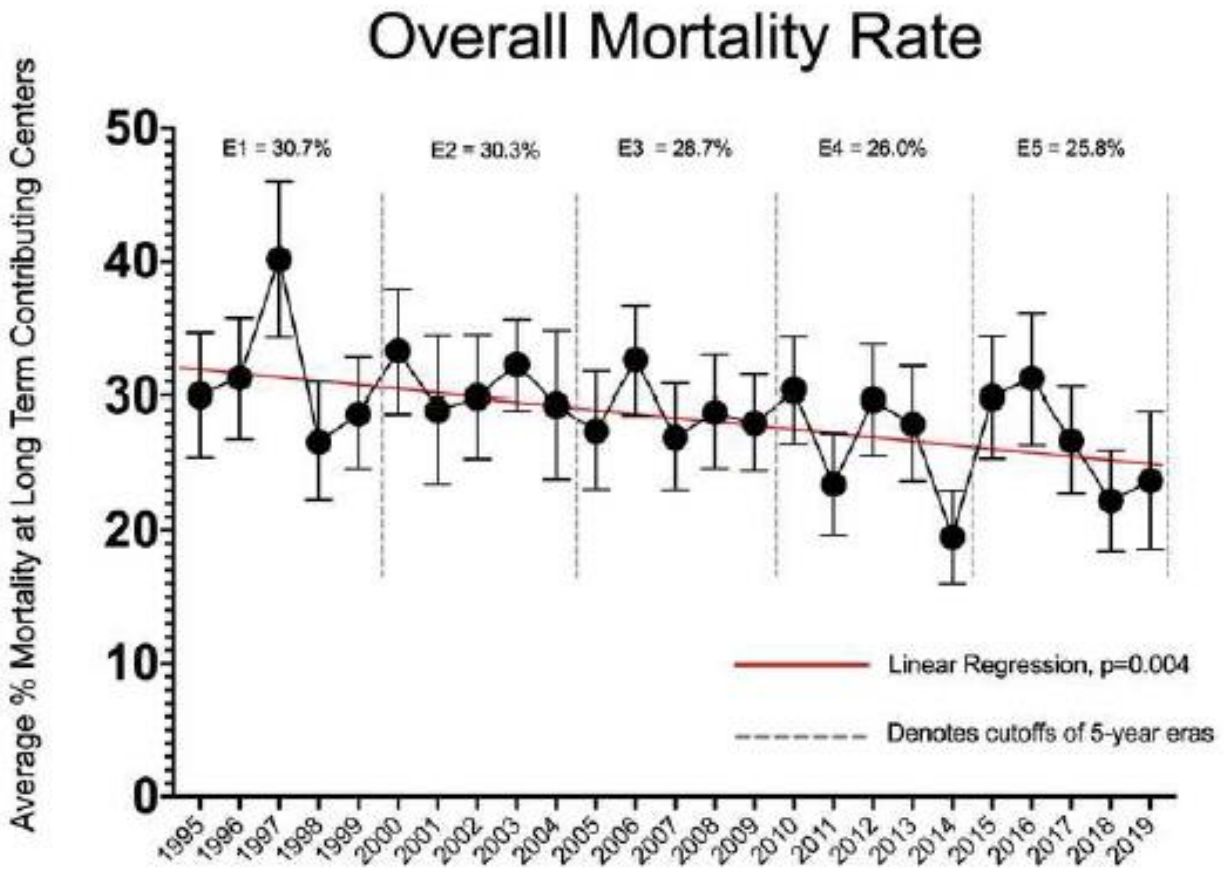
European Reference Network for rare or low prevalence complex diseases

Network Inherited and Congenital Anomalies (ERNICA)

université PARIS-SACLAY

Outcome Improvement

Congenital Diaphragmatic Hernia Study Group
5203 patients over 25 years
Overall mortality 28.2%



Gupta VS et al. Ann Surg, 2021

Pregnancy and chronic disease

1- How will pregnancy affect the patient's disease?

2- How will the presence of a disease alter the course of pregnancy and the health and well being of the fetus?

3- How does the management of the pregnant woman with the particular disease differ from management of the nonpregnant patient?

4- The physician is scared for the fetus and the obstetrician for the affected organs

5- Lack of knowledge of pregnancy physiology leads to unnecessary pregnancy terminations

Diagnosis during pregnancy

Systematic Review

Maternal Bochdalek Hernia during Pregnancy: A Systematic Review of Case Reports

Jin-Young Choi [1,†], Song-Soo Yang [2,†], Jong-Hwa Lee [3], Hyun-Jin Roh [1], Jun-Woo Ahn [1], Jeong-Sook Kim [1], Soo-Jeong Lee [1] and Sang-Hun Lee [1,*]

Diagnostics 2021

- Clinical features of 43 cases of maternal CDH complicating pregnancy
- Asymptomatic to life-threatening complication
- Routine repair for maternal CDH is recommended
  - More symptoms in 2nd or 3rd trimester
  - Risk of visceral strangulation and obstruction
  - Greater visceral displacement in to thorax by enlarging uterus
  - Later stage of pregnancy, operative closure may be difficult

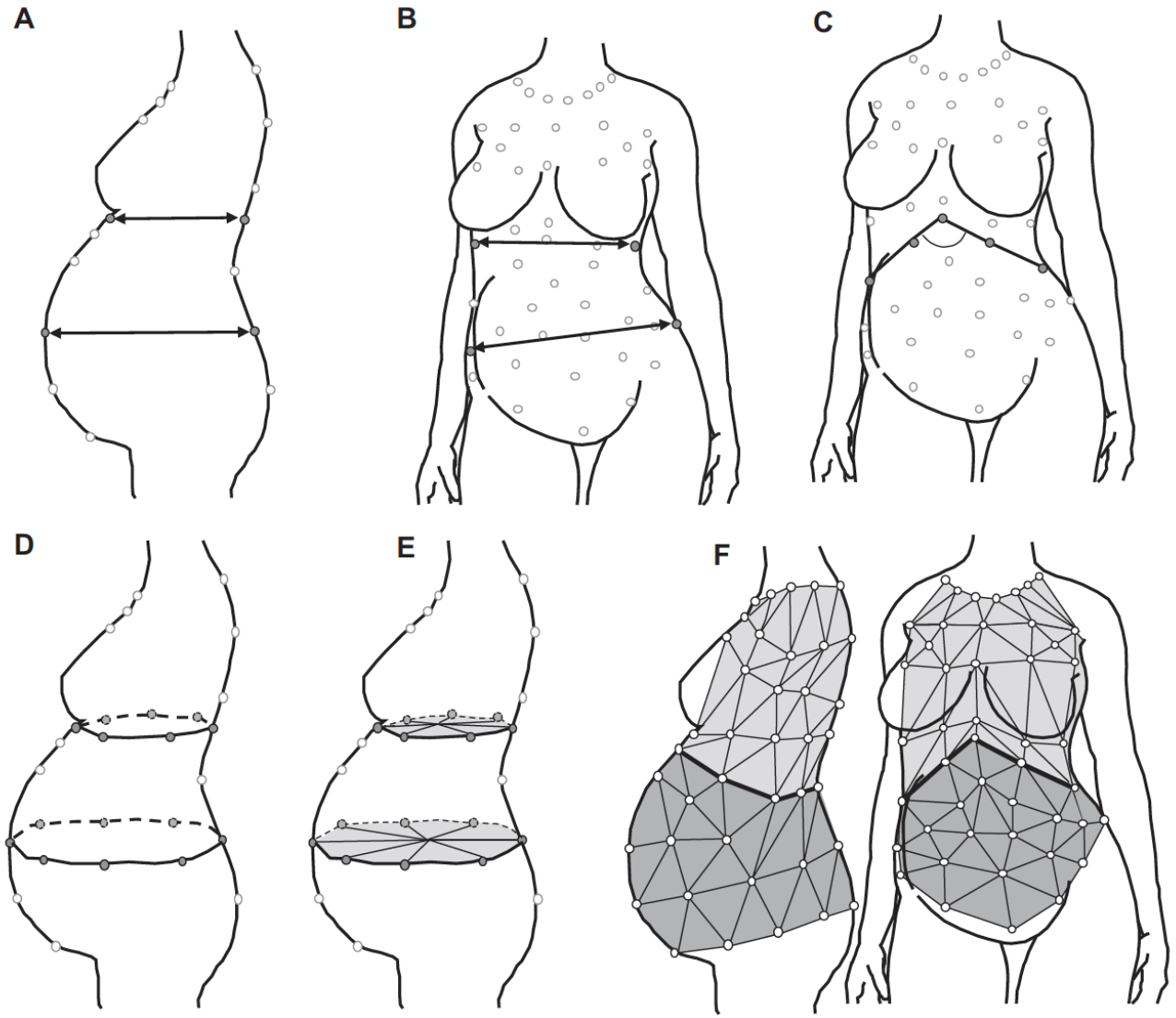
Mean Age (Years)	**28.5**
Gestational age when maternal BH was diagnosed by imaging	
First trimester	1 (2%)
Second trimester	13 (30%)
Third trimester	14 (33%)
Postpartum	15 (35%)
Parity	
Primigravida	19 (44%)
Multiparous	16 (37%)
NA	8 (19%)
Location of maternal BH	
Right side	6 (14%)
Left side	37 (86%)
Hernia defect size (cm)	**5.6**
Number of herniated organs	
1 organ	9 (21%)
2 or 3 organs	18 (42%)
>3 organs	14 (33%)
NA	2 (46%)
Type of herniated organs	
Stomach	27 (63%)
Small bowel	14 (33%)
Colon	30 (70%)
Spleen	9 (21%)
Pancreas	4 (9%)
Omentum	11 (26%)
Cecum	3 (7%)
kidney	1 (2%)
Liver	1 (2%)
Appendix	3 (7%)
Surgical methods	
Laparotomy	19 (44%)
Thoracotomy	10 (23%)
Laparoscopy	5 (12%)
Assisted thoracoscopy	1 (2%)
Laparotomy and thoracotomy	5 (12%)
Non-surgical treatment	3 (7%)
Gestational age at hernia surgery	
Antepartum period	25 (58%)
Postpartum period	15 (35%)
Non-surgical treatment	3 (7%)
Hernia repair methods	
Simple suture	28 (65%)
Suture with mesh	9 (21%)
Mesh only	1 (2%)
No repair	3 (7%)
Unknown	2 (5%)
Mortality	
Fetal/neonatal death	7 (16%)
Maternal death	2 (5%)
Mode of delivery type	
Normal delivery	16 (37%)
Cesarean delivery	20 (47%)
Unknown	7 (16%)
Bowel obstruction, ischemia, perforation of herniated organs	**19 (44%)**
Pregnancy Outcomes	
Preterm birth	15 (35%)
Full term delivery	16 (37%)
Unknown	12 (28%)

65% antenatal

Consequences of physiological changes during pregnancy

- Hormonal
- Cardiovascular
  - 40% increase in cardiac output
    - Increased blood volume and ejection volume
  - Increased myocardial contractility
    - Increased cardiac workload and precarious hemodynamic balance
- Ventilatory function: increased respiratory volumes and hyperventilation
- Musculo-skeletal changes

Adaptation of the thorax during pregnancy

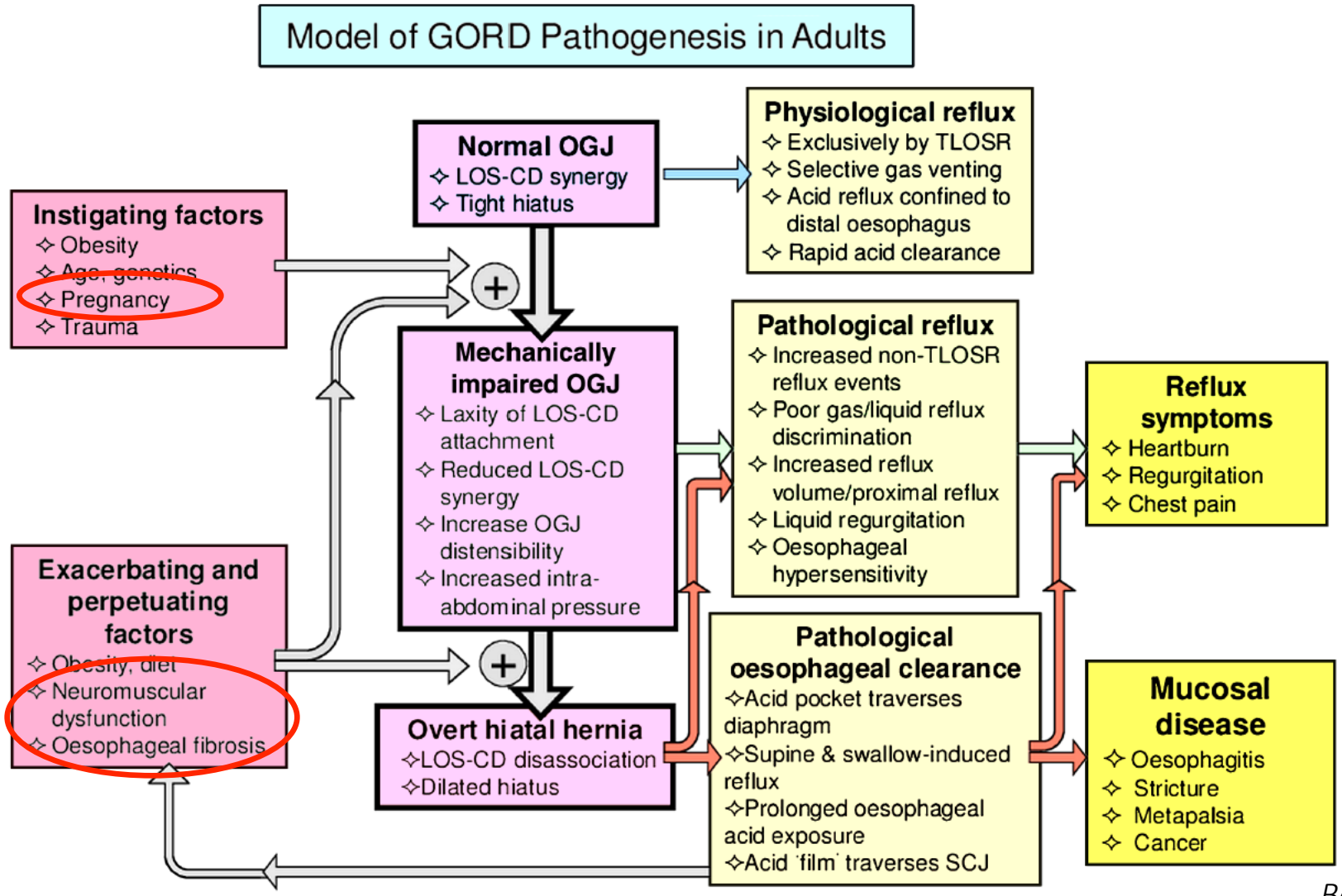


-Chest wall geometry
-Breathing pattern
-Lung and thoracoabdominal volume variations
-Diaphragmatic thickness and motion in seated and supine position
In normal weight range patients

Non invasive techniques
Spirometry by body plethysmography
Optoelectronic plethysmography
US -12-MHz linear probe

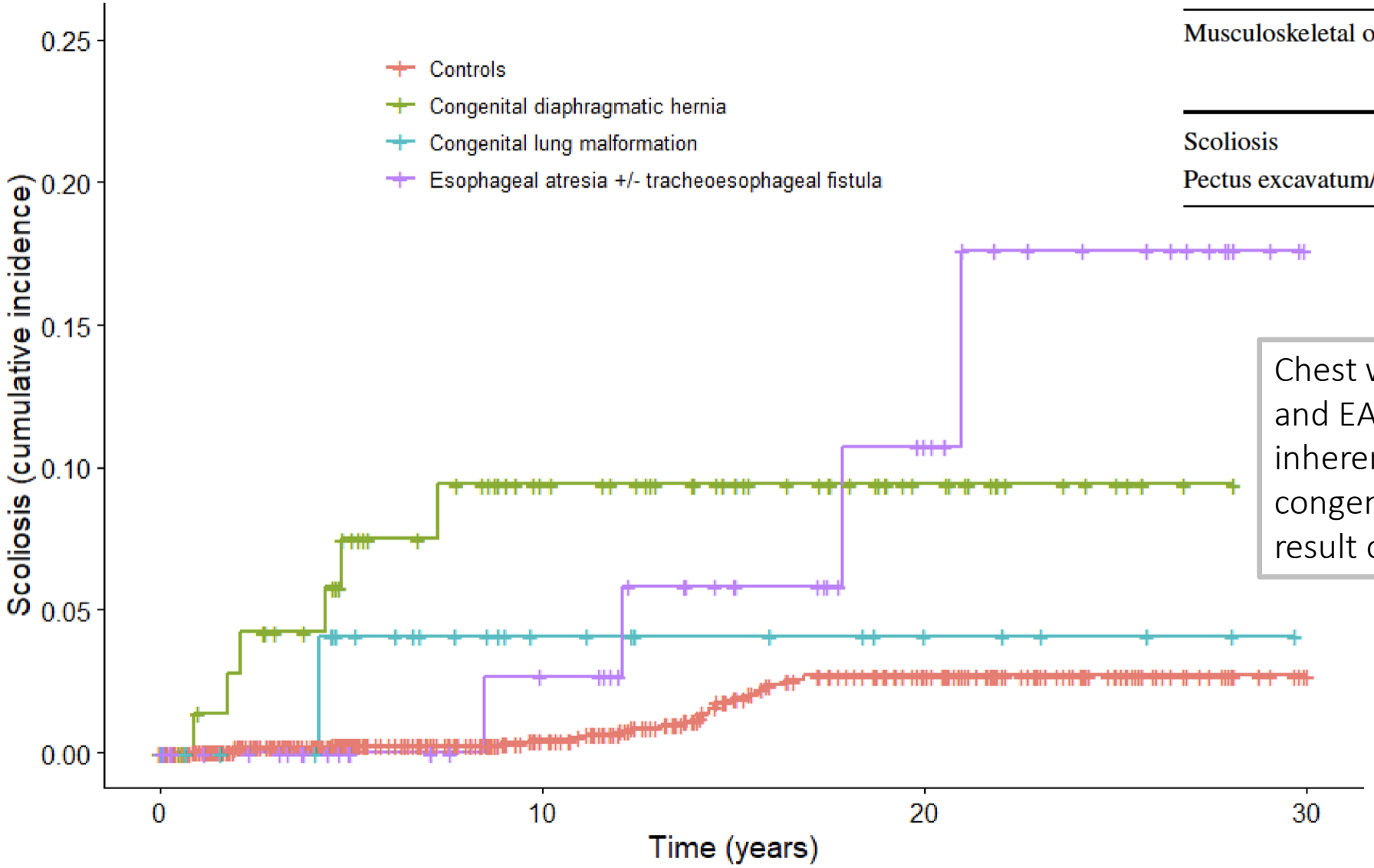
LoMauro A et al. J Appl Physiol 2019

Gastro-oesophageal reflux disease



Boeckxstaens G et al. Gut 2014

Pregnancy and scoliosis



Musculoskeletal outcome	CDH ($n=82$)
	Hazard ratio
Scoliosis	5.03*
Pectus excavatum/ carinatum	1.26

Chest wall anomalies observed in CDH and EA/TEF survivors appeared to be inherently associated with the congenital anomaly and not only a result of the surgical approach

Markel M et al. Ped Surg Int 2022

And for CDH patients?

- Online survey conducted with CDH international
- 34 women born with CDH reported having been pregnant

- 21% (n=7) women had CDH-related issues during pregnancy
  - CDH recurrence
  - Gastro-esophageal reflux
  - Small bowel obstruction
  - Respiratory distress due to chronic restrictive pulmonary disease – termination of pregnancy at 2.5 mo
  - Epidural insertion difficulties due to scoliosis
  - Parietal pain on side of CDH

Median age at survey:

32 years old (IQR: 27-42)

Louise Montalva

And for CDH patients?

	Complications during pregnancy (n=7)	None (n=27)	p
Age (years old)	40	31	0.08
Left side CDH	7/7 (100%)	18/27 (67%)	0.15
Patch repair	2/5 (15%)	11/26 (17%)	0.99
Reintervention for CDH during childhood	0/7 (0%)	8/27 (27%)	0.16
History of scoliosis surgery	3/7 **(43%)**	1/27 **(13%)**	**0.02**

And for CDH patients?

- “Non-severe” CDH neonates (left, no associated malformation, no reintervention for CDH during childhood) may develop complications related to CDH during their adulthood when pregnant
- Scoliosis could be a risk factor for developing complications during pregnancy for CDH survivors
  - High rate of history of scoliosis surgery (43%) vs reported rates of scoliosis between 7-15% in CDH survivors

Conclusion

- Prenatal counselling is needed for CDH patients
- Specific complications can occur and should be anticipated
- In case of IVF procedure, only one embryo should be transferred
- Median age of 40 years in our series / Progress in medical and surgical treatment / More severe cases are surviving
- Open vs thoracoscopic repair:
  - Decrease in small bowel obstruction (Putnam et al. 2017)
  - Decrease in chronic lung disease (Okawada et al. 2021)
  - Decrease in scoliosis rate ?
- International survey in collaboration with CDH International
- International study with OBGYN network