

Prise en charge de la HCD en Chirurgie et Suivi

Quoi neuf dans ce domaine? Retour sur le CDH Symposium Glasgow 2022

Herpie
de **COU**poles
diaphragmatique
Centre de Maladie Rare

Dr Nicoleta Panait

Centre de référence des HCD

Service de chirurgie pédiatrique viscérale

Hôpital Timone Enfants, MARSEILLE

Day 2: Thursday 28th April | 08.30 - 17.45 BST



Lunchtime parallel presentation sessions and Workshop Session -

Oral abstract Presentations ☆

2:50pm - 4:00pm

[View](#)

15:30 - 17:45		Chairs: Dr Irma Capolupo Prof Krisa van Meurs
Session 6: Optimising Intensive and Surgical Care in CDH		
15.30 - 15.45	Standardisation of Care in CDH: are we there yet?	Dr Gregor Walker
15.45 - 15.55	Standardising decision-making in CDH Care: The Japanese Experience	Dr Kouji Nagata
15.55 - 16.15	Update on surgical techniques and materials in CDH	Dr Katrin Zahn
16.15 - 16.35	Approaches to ECLS in CDH	Dr Matthew Harting
16.35 - 17.10	ECLS: My personal philosophy and panel discussion	Dr Kevin Lally, Mr Carl Davis, Prof Dick Tibboel
Oral Abstract Presentations		
17.10 - 17.45	Utility of neurally adjusted ventilatory assist (nava) in weaning and extubation of neonates with congenital diaphragmatic hernia (CDH)	Dr Ana María Sánchez Torres
	Treprostinil in children with congenital diaphragmatic hernia-related pulmonary hypertension	Felix R DeBie
	Heart rate control with landiolol hydrochlorid in CDH infants with ventricular dysfunction	Dr. Lukas Schroeder
	Early pre- and postoperative enteral nutrition in infants with symptomatic congenital diaphragmatic hernia	MD Ulla Lei Larsen
	Anti-reflux surgery in children with congenital diaphragmatic hernia: A prospective cohort study on a controversial practice	Dr Louise Montalva

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Small Bowel Obstruction after neonatal repair of
Congenital Diaphragmatic Hernia – results from a large
longitudinal cohort-study
Katrin Zahn

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L'occlusion intestinale postopératoire résultats de l'équipe de Mannheim

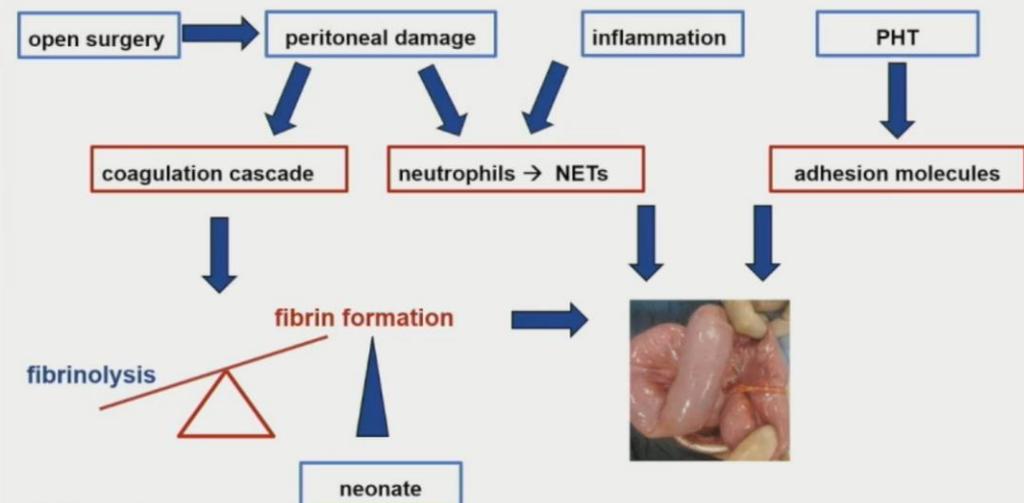
Small bowel obstruction after neonatal repair of CDH
– results from a large longitudinal cohort-study

Katrin B. Zahn, Anna-Maria Franz, Thomas Schaible, Neysan Rafat,
Sylvia Büttner, Michael Boettcher, Lucas M. Wessel

Department of Pediatric Surgery
University-hospital Mannheim, Germany

Dans la littérature récurrences chez 19% des patients avec HCD opérés par voie ouverte versus 2% chirurgie mini-invasive

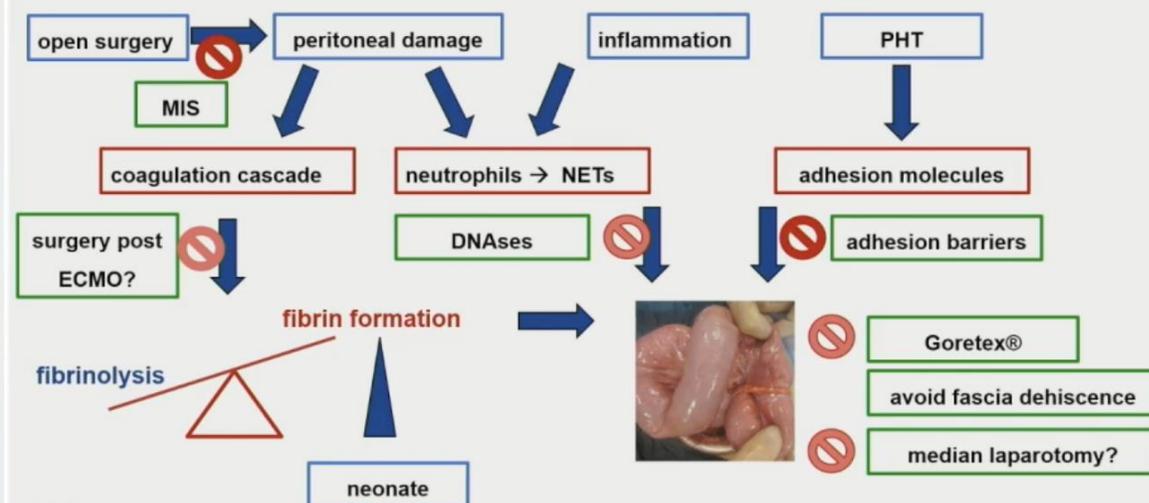
higher risk of SBO in CDH-neonates



Katrin Zahn

Medizinische Fakultät Mannheim
der Universität Heidelberg
Universitätsklinikum Mannheim

higher risk of SBO in CDH-neonates



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der Universität Heidelberg
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Journée Nationale HCD Le Kremlin Bicêtre 20/06/2022



L'occlusion intestinale postopératoire point de vue de l'équipe in Mannheim

Small Bowel Obstruction after neonatal repair of
Congenital Diaphragmatic Hernia – results from a large
longitudinal cohort-study
Katrin Zahn

▪ Conclusions:

Les causes d'occlusion peuvent varier: adhesions, volvulus, récurrence

Risque faible après la chirurgie mini-invasive, mais ne pas libéraliser les indications car ce risque augmente avec le risque de récurrence quand la sélection des patients ne pas rigoureuse

Le type de matériel prothétique (absorbable/non absorbable)

Le type de laparotomie

Nouvelles thérapies pour éviter les adhésions!

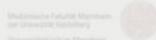
summary SBO after CDH-repair



- different underlying causes → attention: volvulus / recurrence
- CDH-children: high risk for need of surgical intervention (92%)
- significantly lower risk of adhesive SBO after MIS
→ but appropriate patient-selection for MIS and meticulous technique
- longterm results depend on patchmaterial → Goretex® advantageous?
- less peritoneal damage → median laparotomy advantageous ?
- new therapeutic options based on better understanding of underlying cellular and humoral processes



Katrin Zahn



L'occlusion intestinale postopératoire point de vue de l'équipe de Boston

Small Bowel Obstruction after neonatal repair of
Congenital Diaphragmatic Hernia – results from a large
longitudinal cohort-study
Dr Terry Buchmiller

▪ Conclusions

Les anomalies de rotation et fixation
augmentent le risque d'occlusion

En même temps l'absence d'une malrotation
ne protège pas, le risque de volvulus reste
aussi important

Il est important le suivi et bien expliqué les
signes d'occlusion aux parents pour éviter le
retard de présentation à l'hôpital

A bien noter dans le CRO les anomalies de
rotation et fixation et si l'intervention de Ladd a
été réalisée.

Results

Table 2 Bowel status and obstructive complications

Bowel rotation and fixation n=117	Small Bowel Obstruction		Small Bowel Obstruction Requiring Surgical Intervention				
		P		P	Adhesions	Recurrence	Volvulus with SBS
Left CDH no malrotation / fixed	0/1 (0%)	0.999	0/1 (0%)	0.999	0/1 (0%)	0/1 (0%)	0/1 (0%)
Left CDH no malrotation / nonfixed	5/53 (9.4%)	0.193	4/53 (7.6%)	0.254	2/53 (4%)	0/53 (0%)	2/53 (4%)
Left CDH malrotation / nonfixed	10/39 (25.6%)	0.025 *	9/39 (23.1%)	0.014 *	6/39 (15.4%)	1/39 (2.6%)	1/39 (3%)
Right CDH no malrotation / fixed	1/2 (50%)	0.271	1/2 (50%)	0.226	1/2 (50%)	0/2 (0%)	0/2 (0%)
Right CDH no malrotation / nonfixed	0/7 (0%)	0.591	0/7 (0%)	0.596	0/7 (0%)	0/7 (0%)	0/7 (0%)
Right CDH malrotation / nonfixed	1/15 (6.7%)	0.694	0/15 (0%)	0.21	0/15 (0%)	0/15 (0%)	0/15 (0%)

* P<0.05 vs. all other groups

Left CDH with malrotation and nonfixation was a significant predictor for SBO and SBO requiring surgery (P<0.05)
All 3 pts with volvulus had L CDH with nonfixed bowel (100%), however only 1 had malrotation (33%)



Day 3: Friday 29 th April 09.00 - 12.50 BST		
08.30 - 09.00	Registration, tea and coffee	
09.00 - 11.00	Session 7: Transition to Adulthood and longer-term Care	Chair: Dr Hanneke IJsselstijn
09.00 - 09.15	Introduction to the transition to adulthood	Dr Hanneke IJsselstijn
09.15 - 09.35	Clinical Challenges for Adults with CDH	Dr Ulrike Kraemer
09.35 - 09.55	Setting up an Adult CDH Service	Dr Lieke Kamphuis
09.55 - 10.10	Perinatal Care for Women with CDH	Prof Alexandra Benachi
10.10 - 10.45	Oral Abstract Presentations	
	High Altitude Simulation Testing in Patients with Congenital Diaphragmatic Hernia	Dr. Terry L Buchmiller
	Longitudinal evaluation of health status and quality of life in school-aged children with congenital diaphragmatic hernia	Isabel Sreeram
	Pre-school cognitive and social outcomes in CDH survivors	Dr Laura Valfrè
	Self-Reported Respiratory and Gastrointestinal Outcomes in Children Managed at Fetal Treatment Centers with Isolated Congenital Diaphragmatic Hernia	Dr Patrice Eastwood
	Cardio-pulmonary Exercise Testing in Scottish children and adolescents with repaired congenital diaphragmatic hernia	Mr Paul Burns
10.45 - 11.15	Neuropsychological outcome at five years of age in survivors of congenital diaphragmatic hernia	Sophie De Munck De Munck
	Interactive Panel Discussion: Developing improved long-term follow up	Dr Jonathan Coutts

Tester le risque d'hypoxie pendant les vols

Journal of Pediatric Surgery 57 (2022) 195–198

High altitude simulation testing in patients with congenital diaphragmatic hernia

Ashlyn E. Whitlock^{a,*}, Catherine A. Sheils^b, Jill M. Zalieckas^a, Gary Visner^b, Mary Mullen^c, Mollie Studley^a, Lindsay Lemire^a, Terry L. Buchmiller^a

^a Department of Surgery, Boston Children's Hospital, Harvard Medical School, Fegan 3, 300 Longwood Ave, Boston, MA 02115, United States

^b Department of Respiratory Diseases and Pediatrics, Boston Children's Hospital, Harvard Medical School, Boston, MA, United States

^c Department of Cardiology and Pediatrics, Boston Children's Hospital, Harvard Medical School, Boston, MA, United States

Methods- Flight testing

- Altitude is simulated by reducing FIO₂ from 0.21 to 0.15
 - Via nonrebreather facemask, continuous oxygen saturation monitoring (SpO₂)
- Patients initiated testing at baseline (RA or baseline O₂ requirement)
- Testing was considered a “pass” when:
 - SpO₂ maintained > 90%
 - > 94% if diagnosed with pulmonary HTN (RVP>1/2 or on pHTN meds)
- Oxygen was titrated as needed from either room air or their baseline rate of supplementation to achieve a pass
 - If SpO₂ drops below a passing level even with supplementation, the test is concluded, otherwise the test lasts for 15 minutes
 - Average .5L oxygen added, maximum 1 Liter (0.5 liter limit for routine flight)

- Les patients avec HCD risque élevé d'hypoxie pendant les vols
- Malgré la pressurisation dans les vols commerciales, la pression à des altitudes de plus de 10000 mètres est de 565mmHG (N 760 mmHg)
- Test commencé sans ou avec oxygène en fonctions des habitudes
- Il faut que la SpO₂ soit > 90% pour passer le test ou 94% si HTAP

High altitude simulation testing

20 patients testés entre 2006 - 2019.

2 malformations cardiaques

Age moyen 33,4 mois (1 mois et 25 ans).

9 chirurgies avec patch et 6 ECMO

Initial CDH repair and hospitalization of total cohort, those who passed on first attempt and those who eventually passed. $N = 20$ unless otherwise specified.

	Total Cohort ($N = 20$)	Passed on first attempt ($N = 6$)	Eventually Passed ($N = 10$)
Side of defect	15 (75%) left 5 (25%) right	3 (50%) Left 3 (50%) Right	7 (70%) left 3 (30%) right
Size of defect ($n = 18$)	a-3 (16.7%) b-5 (27.8%) c-7 (38.9%) d-3 (16.7%)	a-1 (16.7%) b-3 (50%) c-4 (33.3%) d-2 (0%)	a- 1 (10%) b-3 (30%) c- 4 (40%) d- 2 (20%)
Type of repair: primary vs patch	4 (20%) primary 16 (80%) Patch	2 (33.3%) primary 4 (66.6%) patch	2 (20%) primary 8 (80%) patch
Surgical approach: subcostal vs thoracoscopic	15 (75%) subcostal 5 (25%) thoracoscopic	3 (50%) subcostal, 3 (50%) thoracoscopic	7 (70%) subcostal (3) 30% thoracoscopic
Liver up: ($n = 19$)	9 (42%)	3 (50%)	5 (50%)
Required ECMO	9 (45%)	0	3 (30%)
Required oxygen status at discharge	12 (60%)	3 (50%)	5 (50%)
Required oxygen supplementation at 30 days of age	16 (80%)	3 (50%)	6 (60%)
Discharged on Sildenafil	3 (15%)	11 (16.7%)	3 (30%)
Previous tracheostomy	1 (5%)	0	0

High altitude simulation testing

- 16 patients (80%) ont été sous oxygène à 30 jours et seulement 6 (30%) lors de HAST
- 14 (70%) en air ambiant et 6 (30%) sous oxygène
- 6 (30%) ont passé le test , 1 seul étant sous oxygène
- Aucun patient ayant eu ECMO, defect type D et HTAP

HAST Testing of total cohort, those who passed on first attempt and those who eventually passed. $N = 20$ unless otherwise specified.

	Total cohort ($N = 20$)	Passed on first attempt ($N = 6$)	Eventually passed ($N = 10$)
Pass rate	–	3 (30%)	5 (50%)
Mean age at first test (months)	33.4	5.8	5.6
Mean age at eventual pass (months)	–	5.8	14.7
Attempts to pass	–	1	2.3
Change in SpO ₂ from baseline with FIO ₂ of 0.21–0.15 on initial test ($n = 13$)	8.54	4.3	6.4
Baseline Oxygen requirement at time of test	3 (30%)	1 (16.7%)	3 (30%)
Use of sildenafil at time of test	1 (5%)	1 (16.7%)	1 (10%)
Most recent Echo before test: RVP greater than 1/2	1 (5%)	0	0

Conclusions

Les patients avec HCD peuvent être exposés
à une hypoxie lors des vols

L'ECMO, defect D, persistance HTAP -

Risque élevé d'hypoxie dans des conditions
des pressions basses

Given these findings, it is essential to invest in the education of CDH centers and providers regarding the risks of hypobaric hypoxia and encourage screening of patients who intend to travel in hypobaric environments, especially those with history of severe disease or pulmonary hypertension. Beyond screening those with plans to travel, it may be beneficial to introduce a standard high altitude screening protocol of all patients with CDH and large CDH centers like our own. This would ensure that all at risk patients are properly screened for hypobaric hypoxia and additionally allow for further evaluation of the risk of in-flight hypoxia for patients with CDH.

TRANSITION ENFANT-ADULT

SETTING UP AN ADULT CDH SERVICE

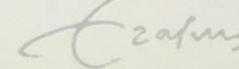


Lieke Kamphuis, MD, PhD
Pulmonologist

Erasmus Medical University Center Rotterdam, The Netherlands

April 29th, 2022

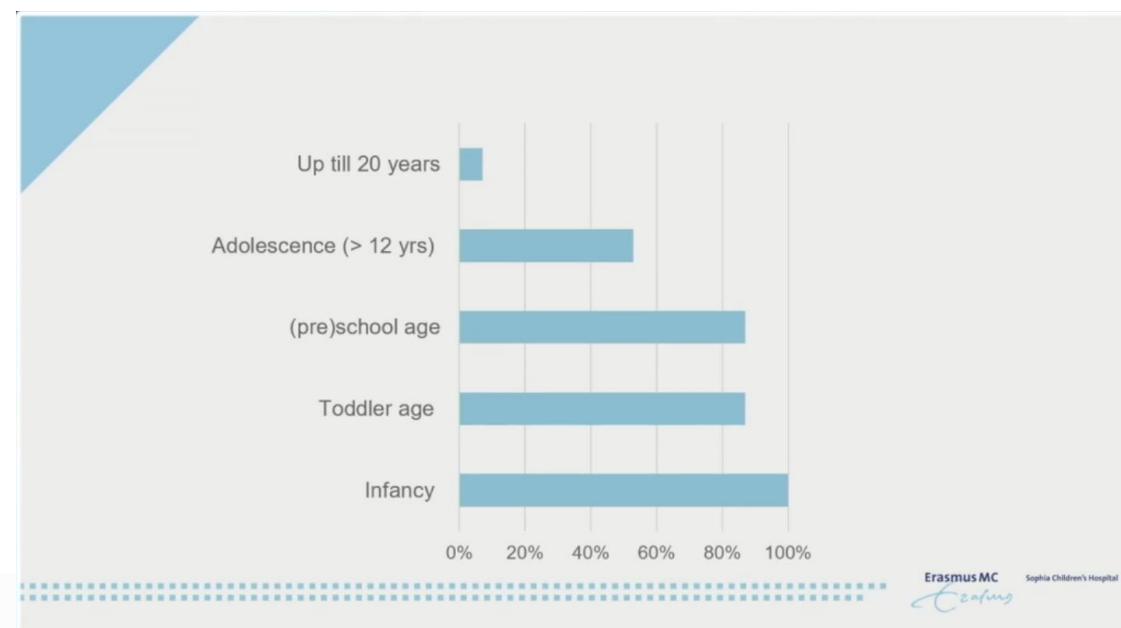
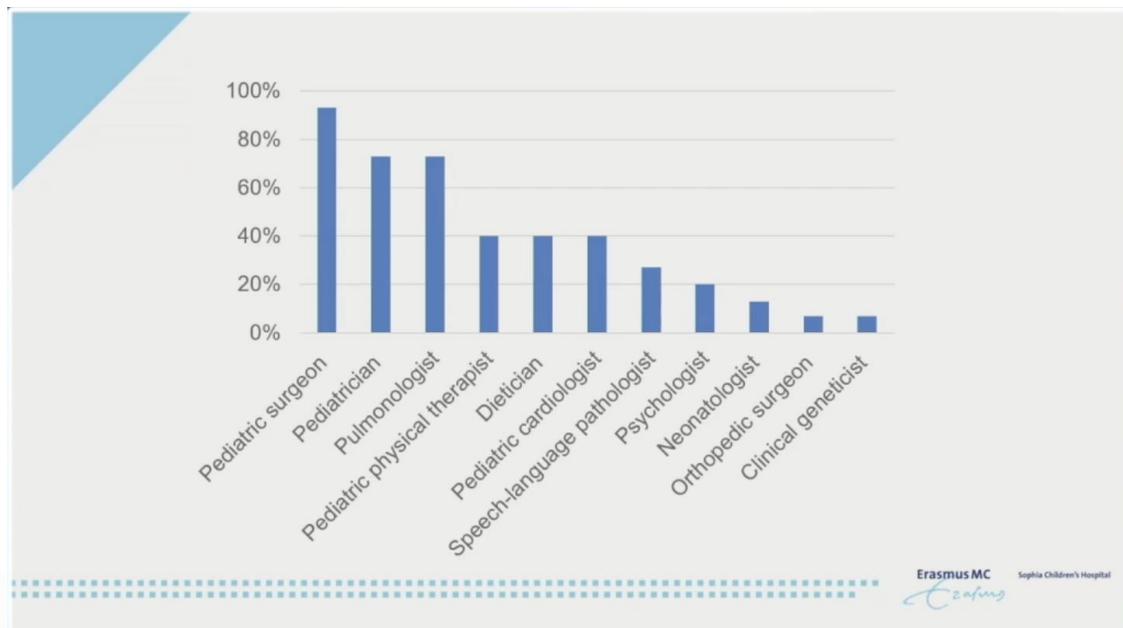
Erasmus MC



Journée Nationale HCD Le Kremlin Bicêtre 20/06/2022

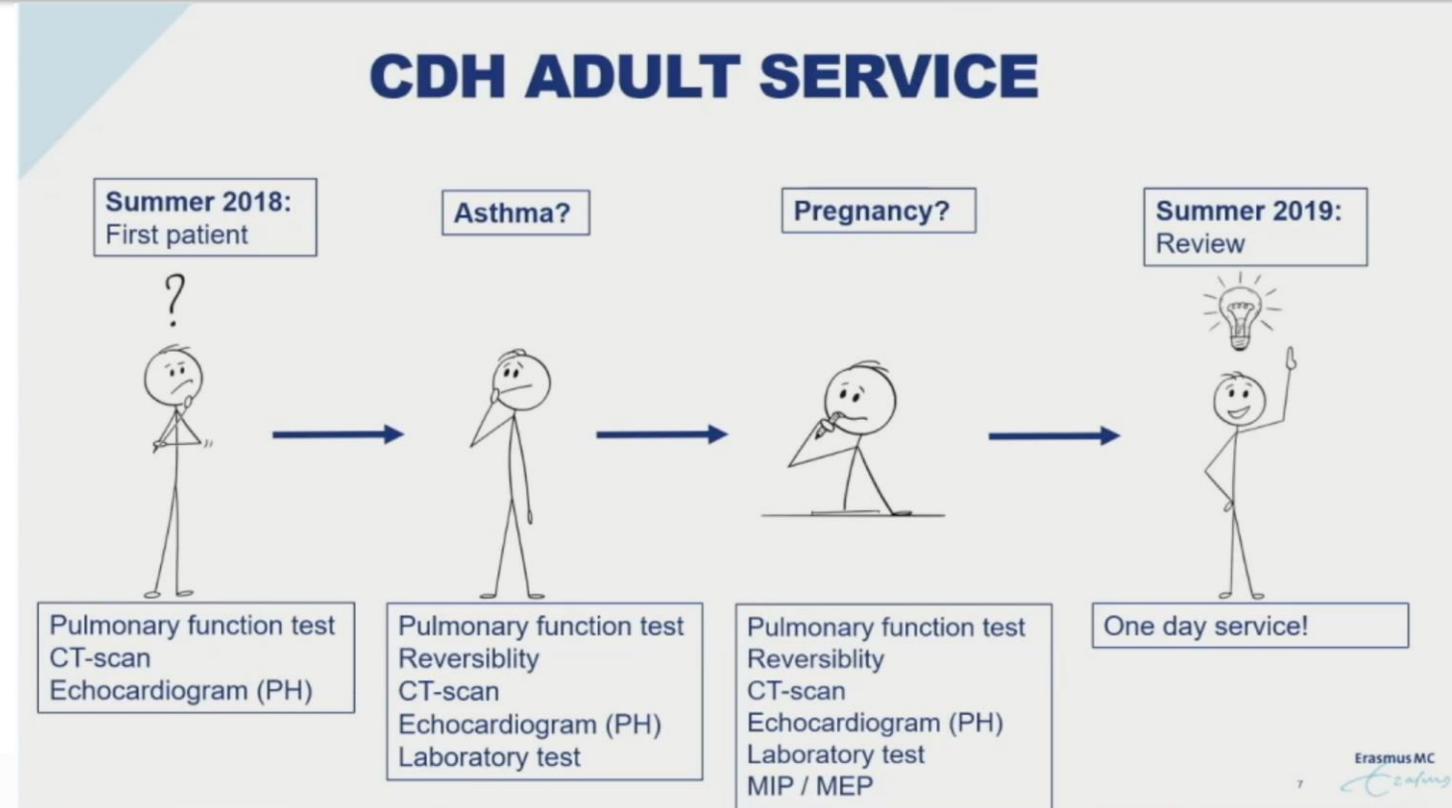


TRANSITION vers les professionnels adultes



TRANSITION vers les professionnels adultes

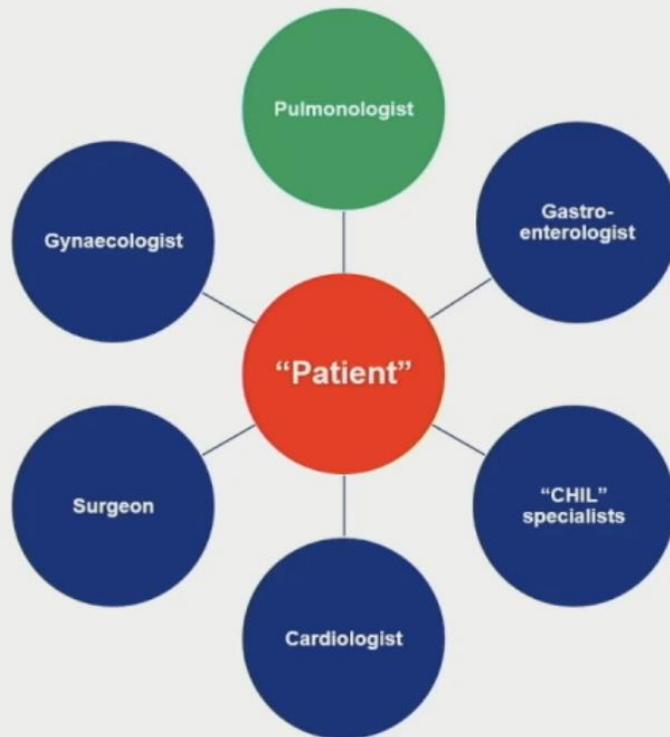
- 92 patients avec HCD depuis 2018
- 5 réopérés pour récurrence avec disparition symptômes respiratoires
- Pas de HTAP
- La grossesse



TRANSITION vers les professionnels adultes

CDH ADULT SERVICE

"Multidisciplinary approach"



- Les symptômes prédominantes sont respiratoires:
Wheezing, difficulté à respirer, toux
Les EFR en faveur d'une obstruction
S'agit-il d'AB?

L'administration des corticoïdes inhalés

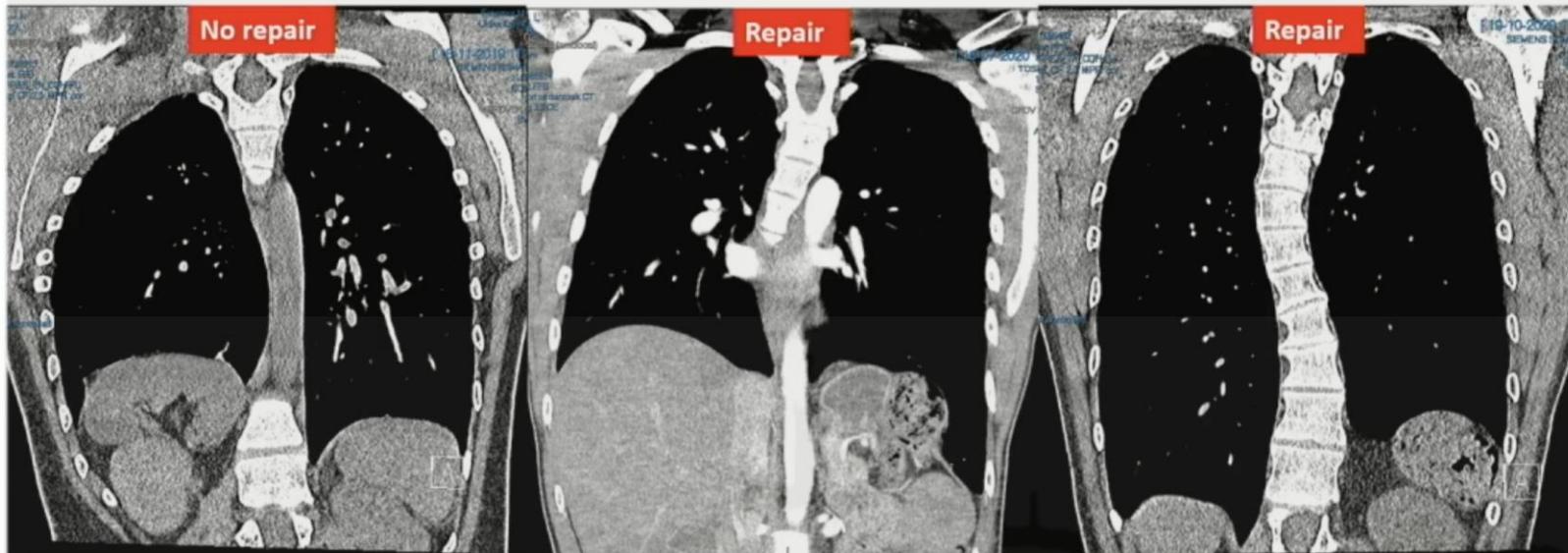
Quand pas d'amélioration il faut considérer ces patients comme emphysème like et les traiter avec beta agonist ou muscarinic antagoniste

La plus part sont bien améliorés Cliniquement et à l'EFR

Récidive à l'âge adulte

- 5 patients ont été opérés de récurrence découverte fortuite grâce à la CS
- La décision d'opérer a été pluridisciplinaire et surtout en fonction des symptômes qui ont disparus après la cire de récurrence

RECURRENCE CDH



Conclusion

- La transition doit être organisée pour éviter que les pédiatres continuent voir ces patients adultes
- Un bilan respiratoire semble le plus important
- Ne pas oublier le scanner thoracique à la recherche de récurrence mais qui donne aussi le diagnostic de l'état de poumon (emphysème?)
- Pour les femmes qui désirent une grossesse faire MIP/MEP

technique of the cone-shaped patch and CDH recurrences: diagnosis

Dr Katrin Zahn, Mannheim Germany

HerDie
de **COU** **pole**
Centre de Maladie Rares
diaphragmatique

*Dr Katrin Zahn,
Département de Chirurgie Pédiatrique
Université Hospital Mannheim, Germany*

Technique of the cone-shaped patch and CDH recurrences: diagnosis

→ CDH-registry (Tsao K et al., 2011):

4390 operated patients (151 thoracoscopic patients)

in-hospital recurrence: **2,7% after open surgery**

Mannheim:

326 patients with completed follow-up >2 yrs.

0,7%

13% cone-shaped patch

9% primary repair

age at recurrence > 12 months:

45%