The long term interdisciplinary follow-up program

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Annual number of CDH patients Rotterdam

Count


19 19 16 17 11 25 24 26 20 21 21 13
Survival after implementation standardized protocol

Van den Hout et al. Fetal Diagn Ther 2011
Annual number of ECMO patients with CDH
Annual survival of CDH patients and ECMO

Year of birth

Survival
- no
- yes

Frequency

Start of Long Term Follow-up 1999

- Patients with severe anatomical congenital malformations
- Patients treated with ECMO

  Added over time:
  - Patients admitted to the ICU after resuscitation
  - Near-drowning patients
  - Traumatic brain injury

- Actual numbers of patients (Oct 2014): 1500
Prospective follow-up schedule

Neonatal

- Months after birth:
  - 6
  - 12
  - 24

Pediatric

- Months after "life-event":
  - 3
  - 12
  - 24

- Age in years:
  - 5
  - 8
  - 12
  - 16
  - 18

Transition
Current disciplines follow-up team

- pediatrician
- pediatric surgeon
- pediatric cardiologist and pulmonologist
- pediatric neurologist
- clinical geneticist

- developmental psychologist
- pediatric physical therapist
- social worker
- ICU nursing staff for 24-hour telephonic helpline
- secretarial office
Long term morbidity

Physical aspects
- cardio-pulmonary morbidity
- gastro-intestinal morbidity
- surgical morbidity
- growth
- motor function development
- exercise tolerance

Psychosocial aspects
- intelligence
- neuro-psychological development
- school functioning
- social-emotional well-being
- behavior
- parental well-being

Quality of Life
## Follow-up program CDH

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<tr>
<th></th>
<th>6 mos</th>
<th>12 mos</th>
<th>24 mos</th>
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## Follow-up program ECMO

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## Follow-up program esophageal atresia

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Follow-up program: tests

- Lung Function at 6 and 12 months and 5, 8, 12 and 18 yrs
- Motor Function assessment
- Exercise capacity assessment
- Psychosocial evaluation
Pulmonary morbidity
Infant Lung Function testing

At 6 – 12 months

Sedation with chloralhydrate 50-75 mg/kg

$V'_{\text{max}}_{FRC}$ : airway obstruction
(Hoo. Am J Respir Crit Care Med 2002)

FRC : lung volume
( Nguyen. Pediatr Pulmonol 2012)
Pulmonary morbidity
Lung Function testing (5) - 8 - 12 - 18 yrs

Dynamic lung volumes: \( \text{FEV}_1, \text{FEV}_{25-75}, \text{FVC}, \text{FEV}_1/\text{FVC} \)

Static lung volumes: TLC, RV, FRC

Diffusion capacity: \( D_{\text{LCO}}, K_{\text{LCO}} \)
(Stam. Pediatr Pulmonol 1996)
Motor function development: Movement Assessment Battery for Children (M-ABC 2)

- manual dexterity
- ball skills
- balance

International used test normative data for children 4-16 years old
Exercise capacity: Bruce protocol

- graded maximum exercise test
  Threadmill; increase speed and slope / 2 min

- criterion of exercise capacity:
  maximal endurance time

- recently established reference data*

* Van der Cammen et al.; Eur J Appl Physiol 2010
Psychosocial aspects (ECMO)

- **6-12-24 mos; 1 hr**
  - BSID II: Baylay Scales of Infant Development II
  - 1-42 mos, mental (verbal / non-verbal) / motoric / behavioural observation scale. Dutch norms

- **5 yrs 1 hr**
  - School?
  - DUX 25: Self-report quality of life (Dutch)
  - PedQl: Pediatric Quality of Life Inventory; 2-18 yrs (international, validated)
  - SDQ: Strengths and Difficulties questionnaire; 3-16 yrs (int. val.)
  - Omitted: RAKIT: IQ
Psychosocial aspects

- 8 yrs; 3 hrs

- DUX 25, PedsQI, SDQ

- CBS-K: experience own competence at school, sport, behaviour, self-esteem (Dutch, validated)

- WISC-III Wechsler Intelligentie Scale for Children - Revised Dutch version. 6-17 yrs; verbal and non-verbal performance scale.

- Bourdon-Vos: attention and concentration, 6-17 yrs
Psychosocial aspects

- 12 yrs; 3 hrs

- DUX 25, PedsQl, SDQ, CBS, WISC III (3 subtests), Bourdon-Vos
- YSR: Young Self-Report, 11-18 yrs. Competence and problem scales
- Tower of London test: executive functioning; planning and solving problems
- 15-wordstest: assessment of memory performance
Conclusions part 1

- Tailormade program
- Model for different patient groups
  - Post cerebral hypoxia
  - Post hypothermia
  - ECMO; neonatal / pediatric
  - Major congenital anomalies
- Patient care; opportunity for early interventions
- Research; prospective, longitudinal data
N= 100; CDH (24), EA (29), SIA (25), AWD (24)

Motor function:
- 17.6% borderline motoric problem, 10.8% definite motor problem
- different distribution than reference population
- bal skills and balance most affected
- CDH and EA most problems

Exercise capacity:
- Mean SDS endurance time -0.5 (SD 1.3), p=0.001
- CDH and EA poorest exercise performance
Cohort Rotterdam and Nijmegen (N=149):

- 13% has severe disability
- Motor function impairment in CDH, PPHN, sepsis, not MAS
- correlation between motor function problems and cognitive impairment
Deterioration of exercise capacity after neonatal extracorporeal membrane oxygenation

M.H.M. van der Cammen-van Zijp*#, S.J. Gischler*#, W.C.J. Hop*, J.C. de Jongste*, D. Tibboel*# and H. IJsselstijn*#

N=120

5 – 8 – 12 yrs

Exercise capacity: Bruce protocol

CDH 15%     MAS 60%     other 25%
Maximal exercise performance 5 - 8 - 12 yrs

Data shown are ANOVA estimates of mean values with 95% confidence intervals. Squares: CDH  asterisks: MAS  triangles: remaining group  circles: total group

van der Cammen-van Zijp. Eur Respir J 2011
Prospective longitudinal evaluation of lung function during the first year of life after repair of congenital diaphragmatic hernia

Marjolein Spoel, MD; Lieke van den Hout, MD; Saskia J. Gischler, MD, PhD; Wim C. J. Hop, PhD; Irwin Reiss, MD, PhD; Dick Tibboel, MD, PhD; Johan C. de Jongste, MD, PhD; Hanneke IJsselstijn, MD, PhD

- N= 37
- 32% ECMO 41% O₂ day 28, CLD / BPD
- Lung function 6 – 12 mos
- FRCₚ: lung volume 13-26 ml/kg, >26/ml = hyperinflation
CDH: Lung function first year of life


ECMO-BPD
ECMO-noBPD
noECMO-BPD
noECMO-noBPD

FRC (ml/kg)

33.9 (1.8)  29.8 (2.3)  31.5 (1.6)  25.6 (1.1)

*
Diagnosis-related deterioration of lung function after extracorporeal membrane oxygenation

Marjolein Spoel, Roxanne Laas, Saskia J. Gischler, Wim J.C. Hop, Dick Tibboel, Johan C. de Jongste and Hanneke IJssestijn

- N=121
- Lung Function at 5 – 8 – 12 yrs
- Duration of ventilation / ECMO
  → neg influence FEV\textsubscript{1} and FVC
Figure 3. SDS FEV$_1$ (mean, 95%CI) after BD at 5, 8 and 12 years for the different subgroups.
Physical growth in neonatal ECMO: height

![Graph showing height in ECMO with SDS and time points such as 0.5 yr, 1 yr, 2 yrs, 5 yrs, 8 yrs, and 12 yrs. Symbols and error bars indicate comparisons for CDH, MAS, and Other groups. Statistical significance levels are marked with + for p < 0.05, † for p < 0.01, and ‡ for p < 0.001.]
Physical growth in neonatal ECMO: weight

Weight in ECMO

-3 -2 -1 0 1

MAS CDH 0,5 yr 1 yr 2 yrs 5 yrs 8 yrs 12 yrs

Other

+ p < 0,05 † p < 0,01 ‡ p < 0,001
- N= 35, 8 yrs

- 16 ECMO   19 non-ECMO

- Mental development assessment

- Motor function assessment
Intelligence at 8 years in CDH patients

Black: IQ < -1 SD; white: average IQ (between -1 and +1 SD); dashed: IQ > +1SD

Madderom et al., Arch Dis Child Fetal Neonat, 2013
Motor function at 8 yrs in CDH patients

Figure 3  Motor function in 8-year-old CDH patients. In black, severe motor function problems; in grey, borderline motor function problems; in white, normal motor function. Number of patients is indicated in the bars. CDH, congenital diaphragmatic hernia; ECMO, extracorporeal membrane oxygenation; Non-ECMO, no extracorporeal membrane oxygenation. TIS, total impairment score.

Madderom et al., Arch Dis Child Fetal Neonat, in press
Combined intelligence and motor function at 8 years in CDH

- black: IQ en motor function normal
- white: IQ en motor function both abnormally low
- grey: IQ abnormally low only

ECMO-CDH
- 31%
- 46%
- 23%

nonECMO-CDH
- 8%
- 8%
- 85%

Madderom et al., Arch Dis Child Fetal Neonat, 2013
Concentration at 8 yrs in CDH

- 68% low-very low information processing speed
- 36% low-very low accuracy
- Feeling of self-competence: well developed (PedQI)

Madderom et al., Arch Dis Child Fetal Neonat, 2013
### School performance at 8 years in CDH

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<th>ECMO n=16</th>
<th>Non-ECMO n=19</th>
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<tr>
<td>Regular education</td>
<td>8 (50%)</td>
<td>11 (58%)</td>
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<tr>
<td>Regular education with extra help</td>
<td>7 (44%)</td>
<td>7 (37%)</td>
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<tr>
<td>Special education</td>
<td>1 (6%)</td>
<td>1 (5%)</td>
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CDH at 8 yrs

- CDH patients at risk for long-term morbidity
- especially motor-function and concentration
Sensorineural hearing loss and language development following neonatal extracorporeal membrane oxygenation.


- N=136
- 5 yrs
- >20 dB significant hearing loss, sensorineural or conductive

→ 3.7% bilateral sensorineural hearing loss: higher prevalence than normal population

→ normal language development and IQ
Conclusions part 2

Following CDH children are at risk for:

- Delayed motor function performance
- Decreased exercise tolerance
- Increasing airflow obstruction
- Problems with school performance despite normal intelligence
# The Interdisciplinary Follow-up Team for Patients and Parents

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<th>Clinical geneticist</th>
<th>Pediatric surgeons</th>
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Research: PhD-theses on follow-up