UMC Utrecht

Aangeboren hartafwijking
Denk aan de hersenen

Hans Breur
Kindercardioloog

CarVasZ 2017

Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE

Disclosure belangen spreker

(potentiële) belangenverstrengeling	Geen / Zie hieronder
Voor bijeenkomst mogelijk relevante relaties met bedrijven	Bedrijfsnamen
• Sponsoring of **onderzoeksgeld** • Honorarium of andere (financiële) vergoeding • Aandeelhouder • Andere relatie, namelijk …	• • • stichting hartekind

NVHVV Nederlandse Vereniging voor Hart en Vaat Verpleegkundigen

Overleving met een congenitale hartafwijking

Van der Linde, J Am Coll Cardiol 2011

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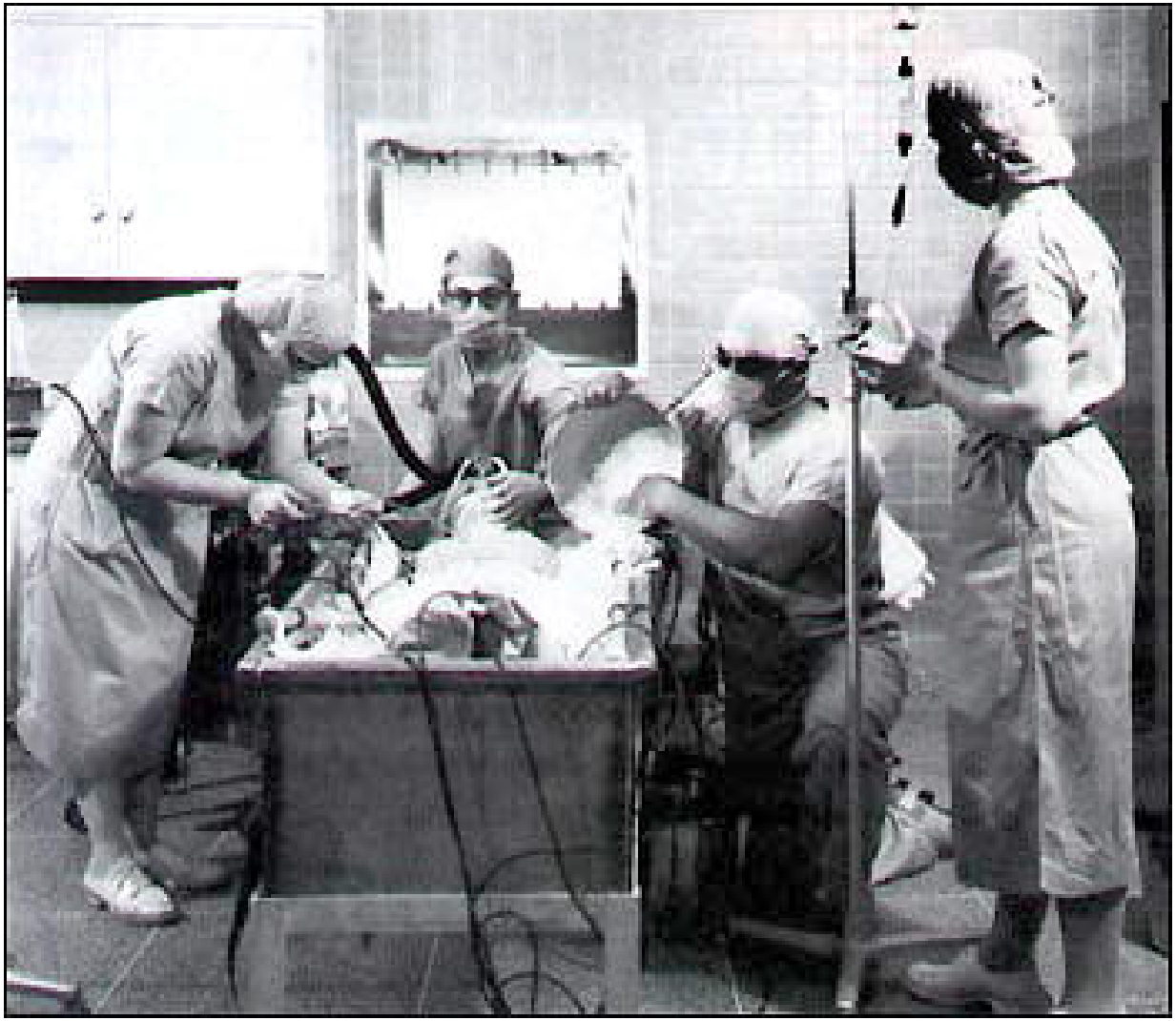
PREVENTING COLLATERAL DAMAGE

Overleving met een congenitale hartafwijking



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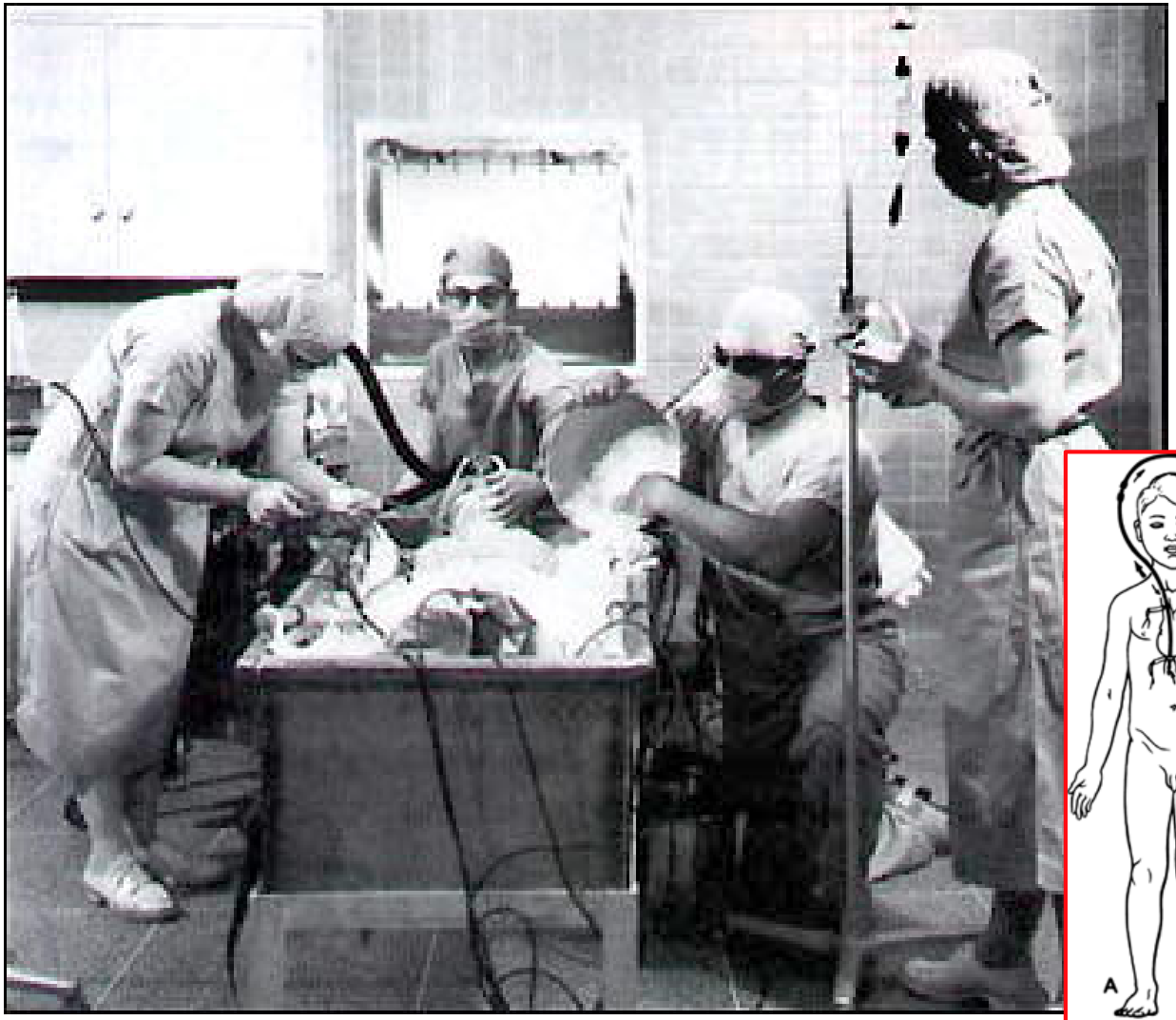
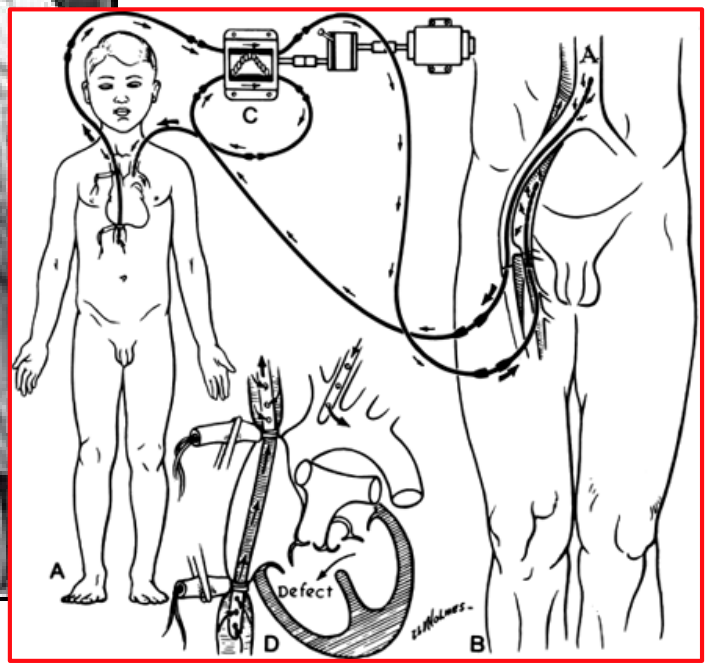
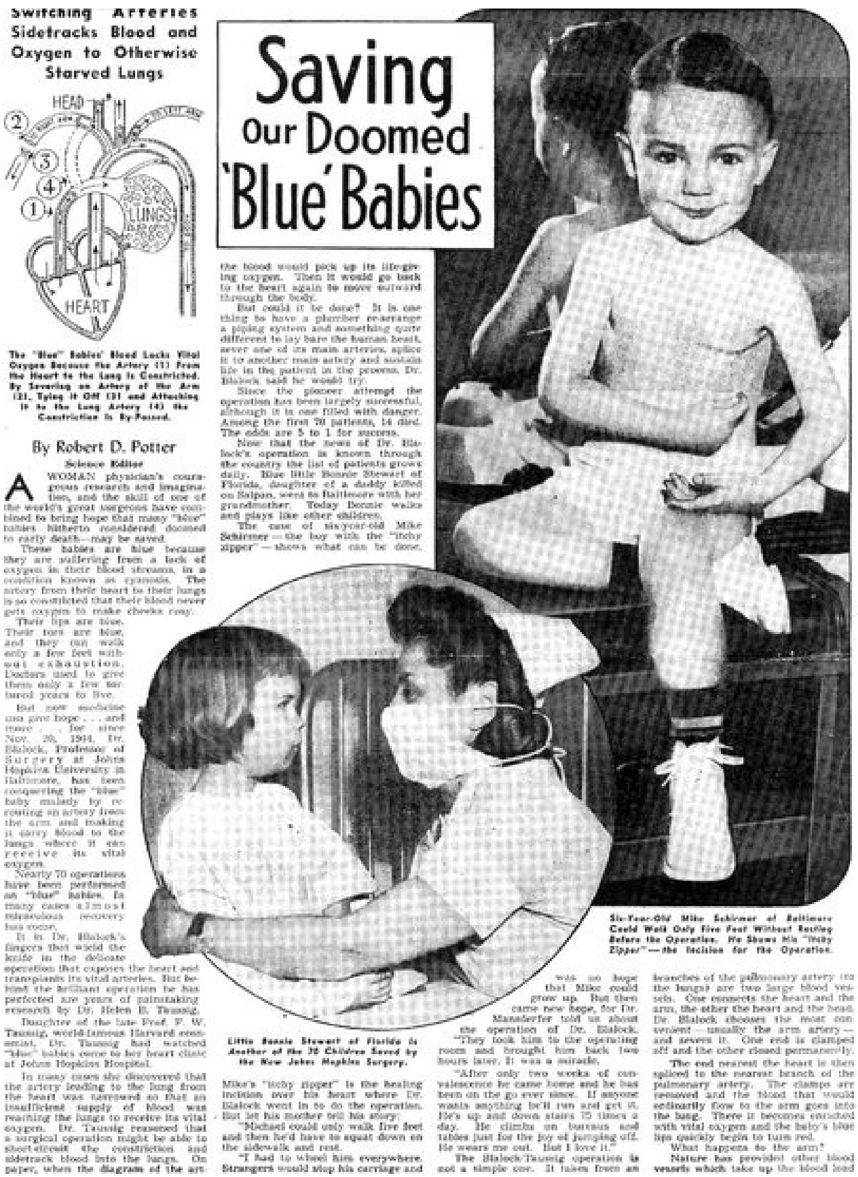





gettyimages
Bettmann
515137782

Overleving met een congenitale hartafwijking


Switching Arteries Sidetracks Blood and Oxygen to Otherwise Starved Lungs

The "Blue" Babies' Blood Lacks Vital Oxygen Because the Artery (1) From the Heart to the Lung Is Constricted. By Severing an Artery of the Arm (2), Tying It Off (3) and Attaching It to the Lung Artery (4) the Constriction Is By-Passed.

Saving Our Doomed 'Blue' Babies

By Robert D. Potter
Science Editor

A WOMAN physician's courageous research and imagination, and the skill of one of the world's great surgeons have combined to bring hope that many "blue" babies hitherto considered doomed to early death—may be saved.

These babies are blue because they are suffering from a lack of oxygen in their blood streams, in a condition known as cyanosis. The artery from their heart to their lungs is so constricted that their blood never gets oxygen to make cheeks rosy.

Their lips are blue. Their toes are blue, and they can walk only a few feet without exhaustion. Doctors used to give them only a few tortured years to live.

...

the blood would pick up its life-giving oxygen. Then it would go back to the heart again to move outward through the body.

But could it be done? It is one thing to have a plumber rearrange a piping system and something quite different to lay bare the human heart, sever one of its main arteries, splice it to another main artery and sustain life in the patient in the process. Dr. Blalock said he would try.

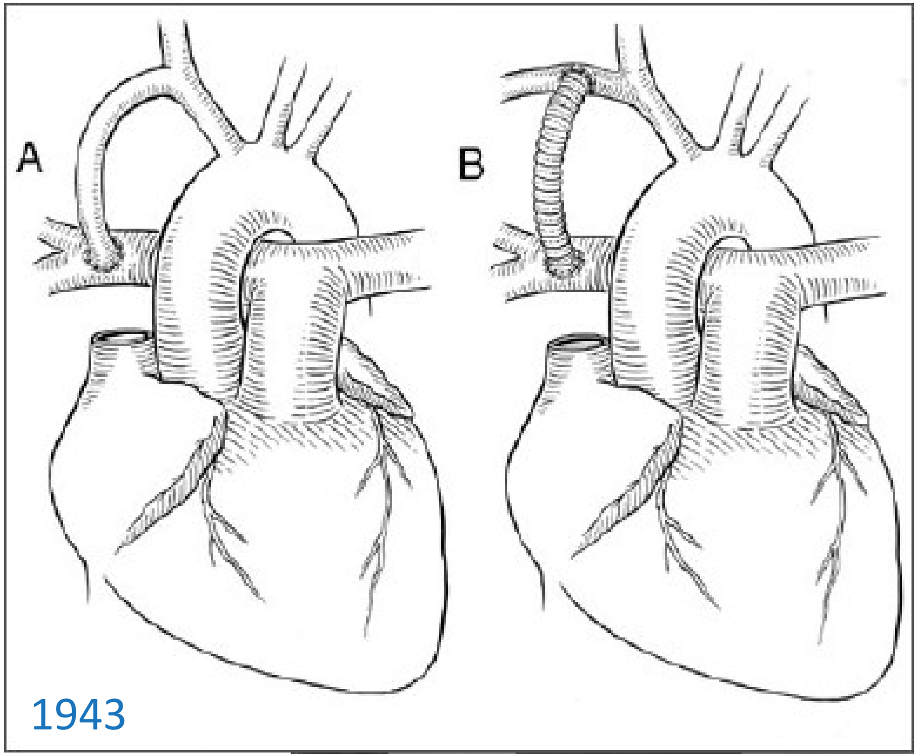
Since the pioneer attempt the operation has been largely successful, although it is one filled with danger. Among the first 70 patients, 14 died. The odds are 5 to 1 for success.

Now that the news of Dr. Blalock's operation is known through the country the list of patients grows daily. Blue little Bonnie Stewart of Florida, daughter of a daddy killed on Saipan, went to Baltimore with her grandmother. Today Bonnie walks and plays like other children.

The case of six-year-old Mike Schirmer — the boy with the "itchy zipper" — shows what can be done.

Six-Year-Old Mike Schirmer of Baltimore Could Walk Only Five Feet Without Resting Before the Operation. He Shows His "Itchy Zipper" — the Incision for the Operation.

Little Bonnie Stewart of Florida Is Another of the 70 Children Saved by the New Johns Hopkins Surgery.



Overleving met een congenitale hartafwijking

1953
ASD sluiting

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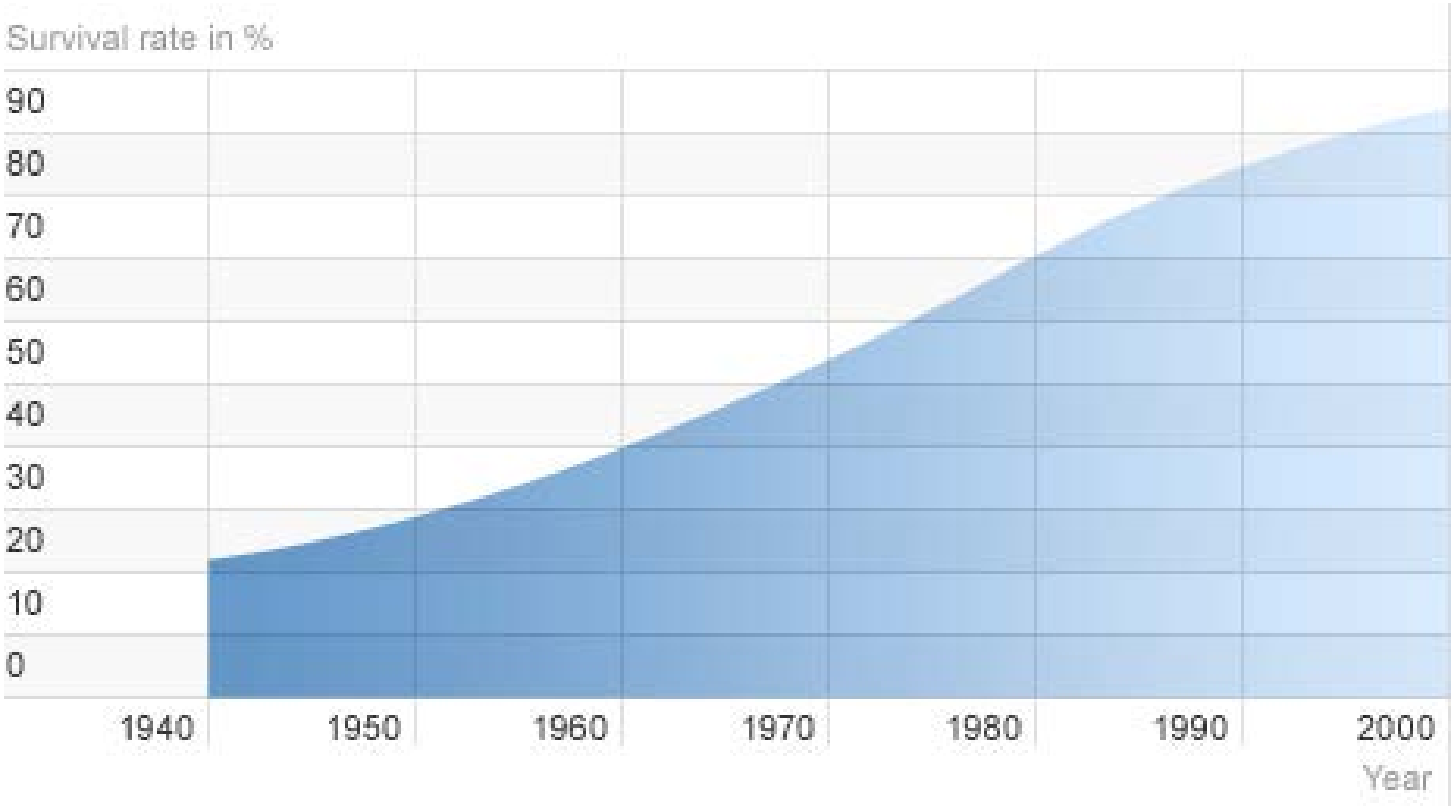
Overleving met een congenitale hartafwijking



Van der Linde, J Am Coll Cardiol 2011

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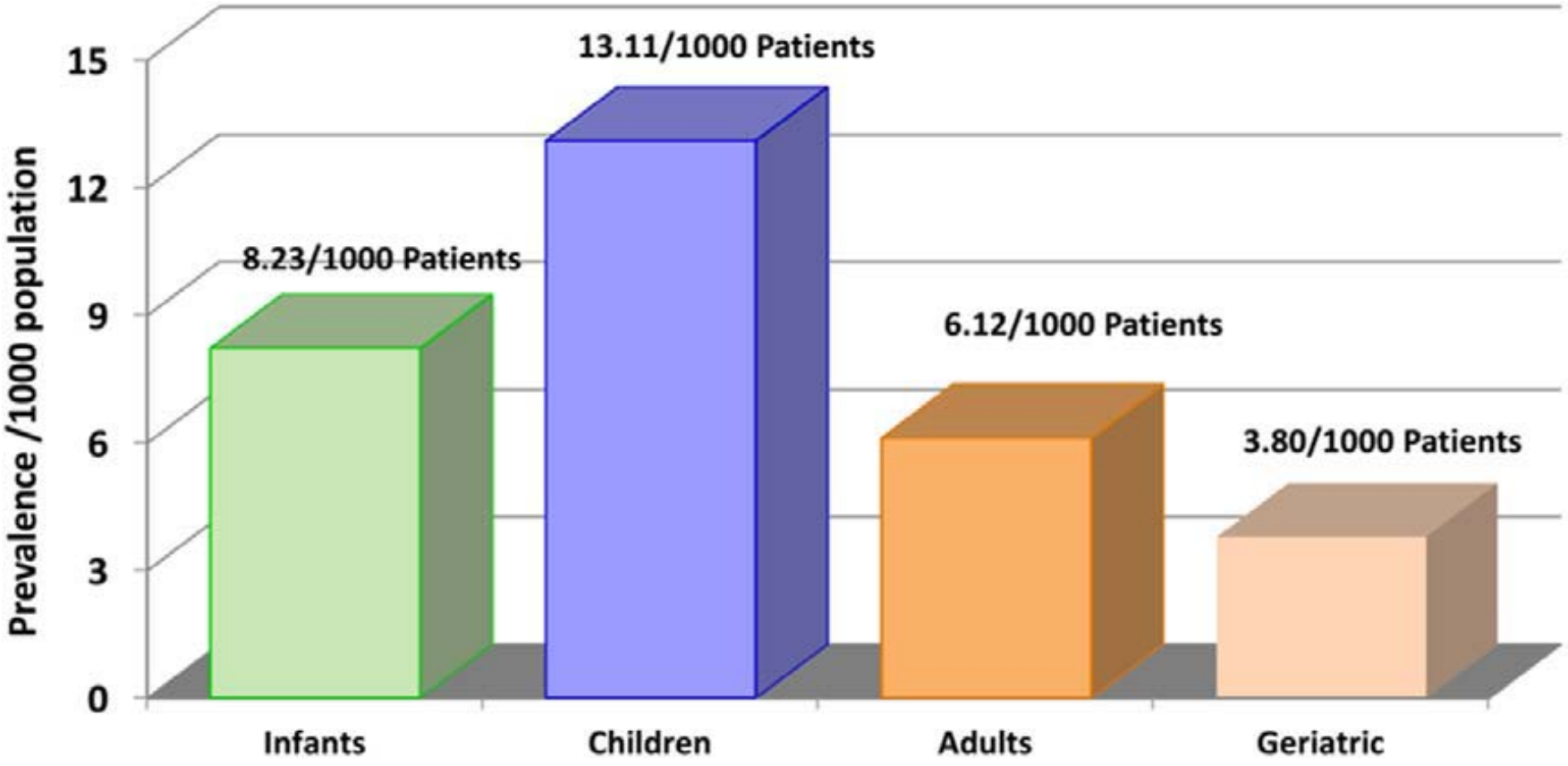
PREVENTING COLLATERAL DAMAGE

Overleving met een congenitale hartafwijking



Mazor Dray & Marelli, 2015.

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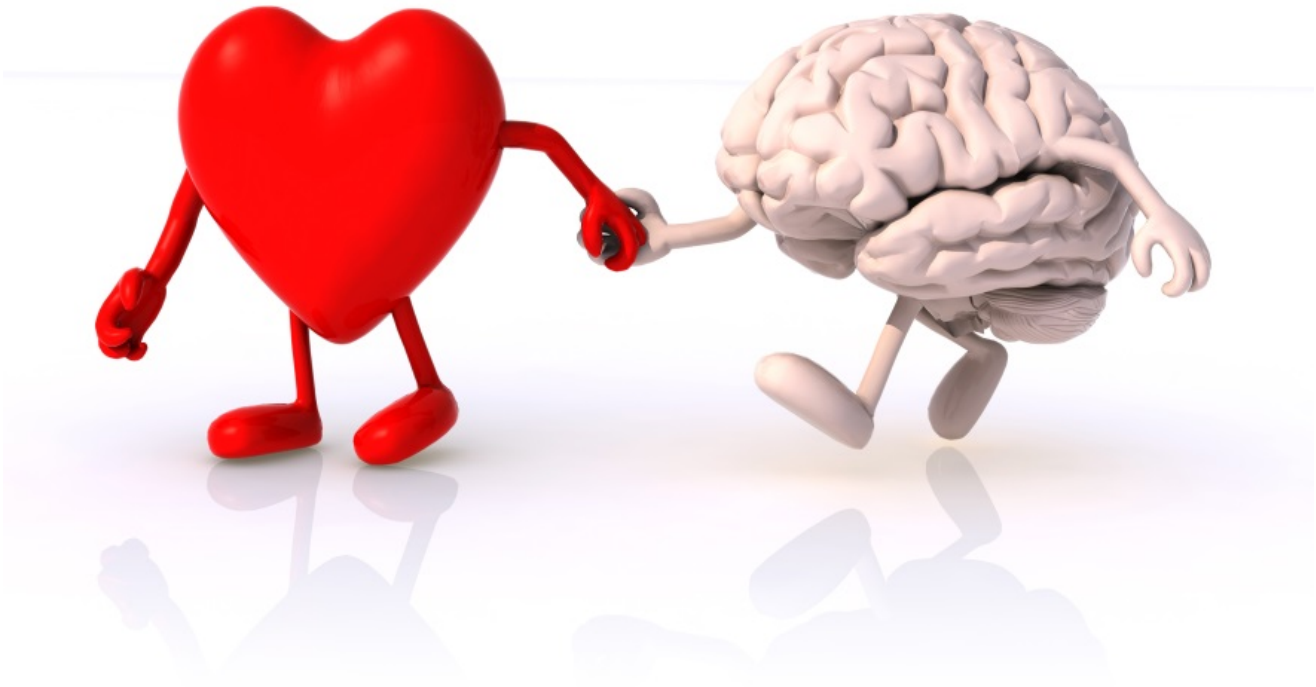
Congenital Heart Disease Life Span

PREVENTING COLLATERAL DAMAGE



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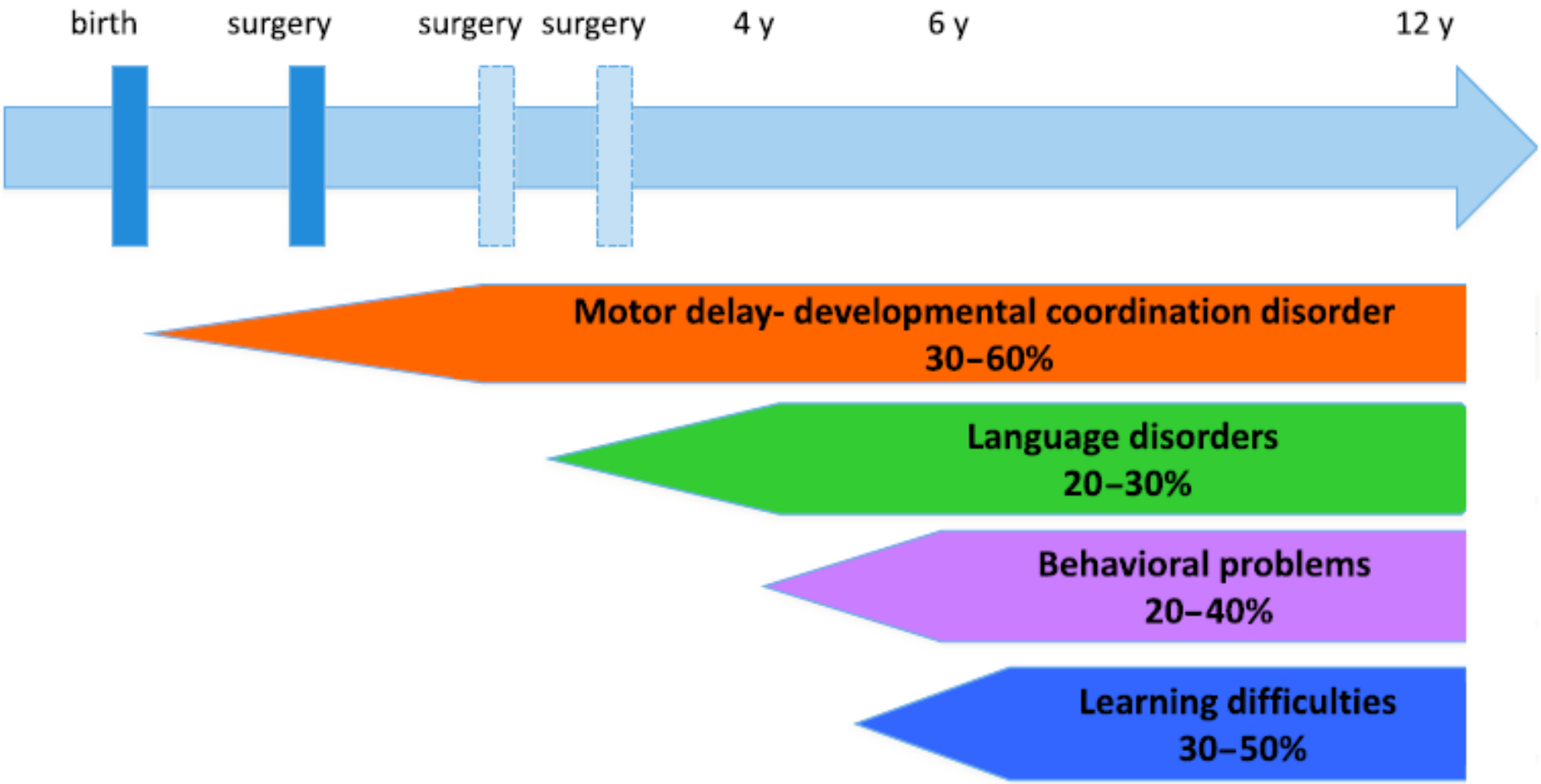
PREVENTING COLLATERAL DAMAGE



Latal, Clinics in Perinatology, 2016.

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PREVENTING COLLATERAL DAMAGE

Cognitieve ontwikkeling op schoolleeftijd

Ontwikkeling	% van de kinderen waarin afwijkend
Zelfredzaamheid	22%
Sociale participatie	13%
Communicatie	17%
Aanpassingsvermogen	15%

Majnemer, Jped 2008

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PREVENTING COLLATERAL DAMAGE

Neurologische ontwikkeling in adolescenten

	Patients (n=59), mean (SD)	Comparison children (n=40), mean (SD)	p
Wechsler Intelligence scale for children			
Full scale IQ	103.10 (16.49)	112.68 (10.43)	0.001
Verbal comprehension	109.14 (18.83)	115.21 (15.96)	0.11
Perceptual reasoning	103.66 (14.77)	113.82 (8.83)	≤0.001
Working memory	93.42 (13.64)	103.76 (12.19)	≤0.001
Processing speed	101.46 (14.19)	105.39 (11.20)	0.15

Schaefer, dev med child neurol 2013

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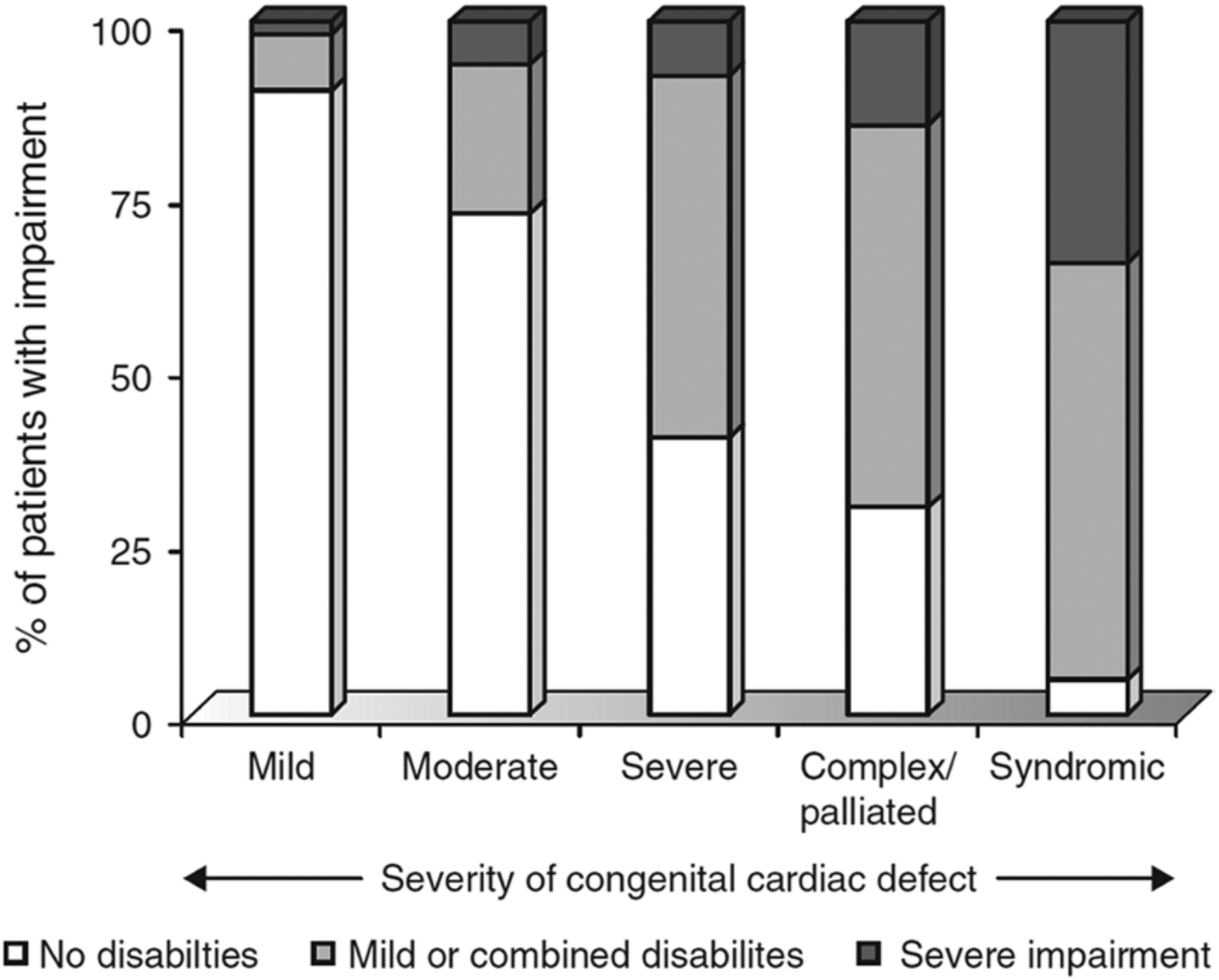
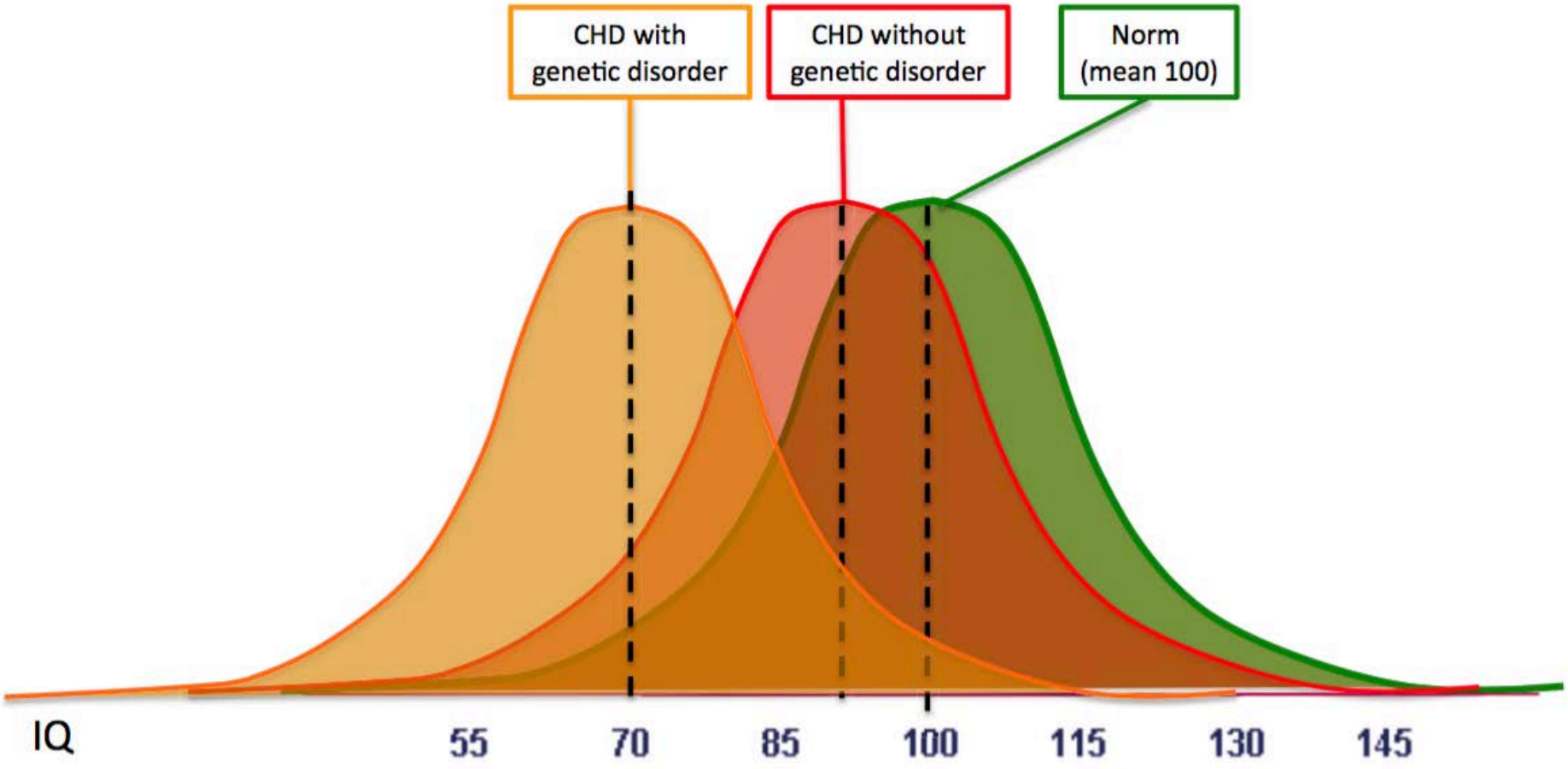
Wernovsky, Cardiol Young 2006.



Cognitieve ontwikkeling



Latal, Clinics in Perinatology, 2016.

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Andere populaties met neonatale problemen

Table 1
Overview of major outcome categories in the largest at-risk population

	HIE	Very Preterms	CHD
Prevalence	1–6/1000	10/1000	6/1000
Cerebral palsy	30% TH: 20%	5%–10%	2%
IQ <70	30%	10%–20%	10%–20%
Mild deficits	~50%	~50%	~30%–50%

Abbreviations: HIE, hypoxic-ischemic encephalopathy; TH, therapeutic hypothermia.

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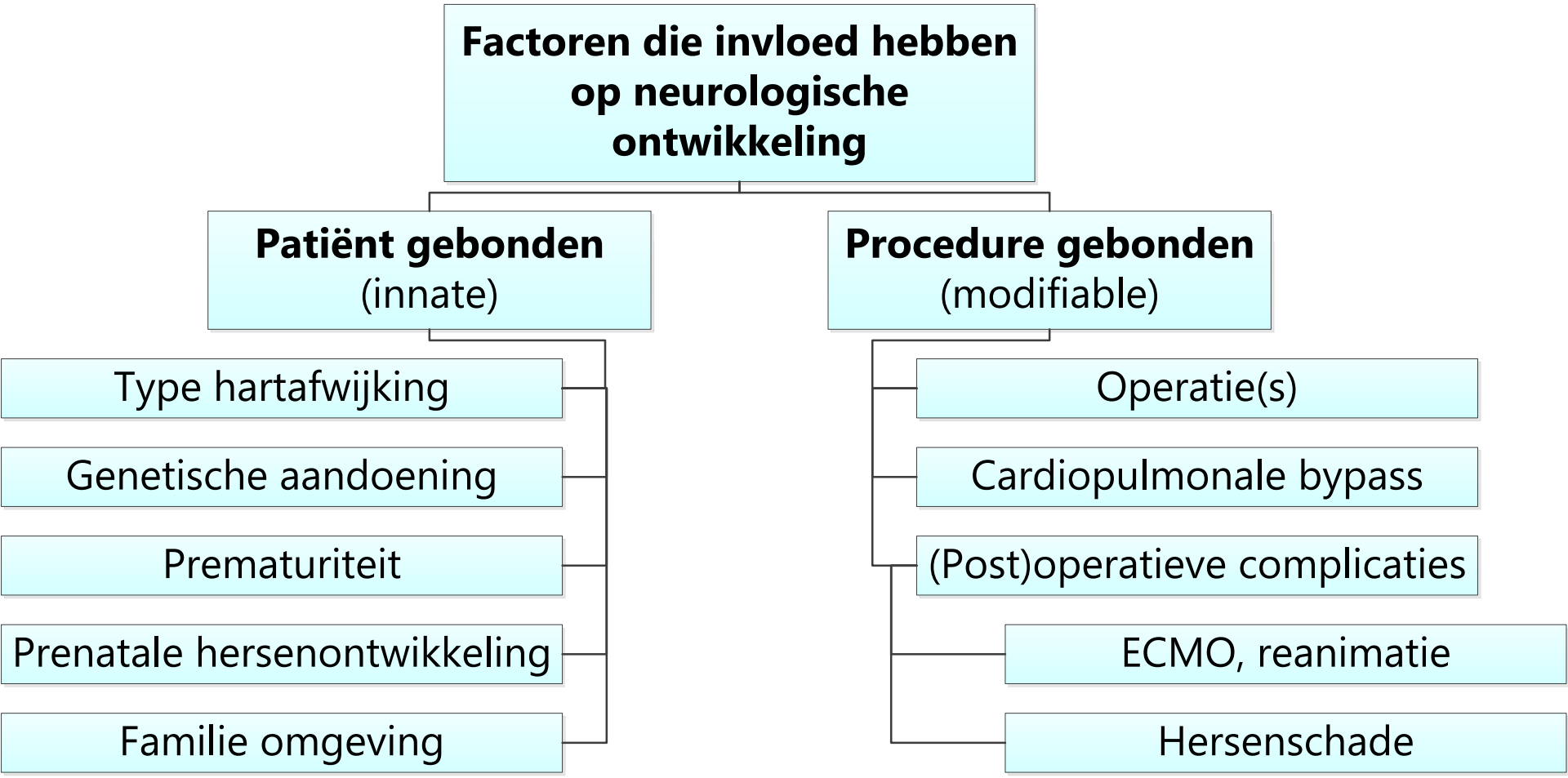
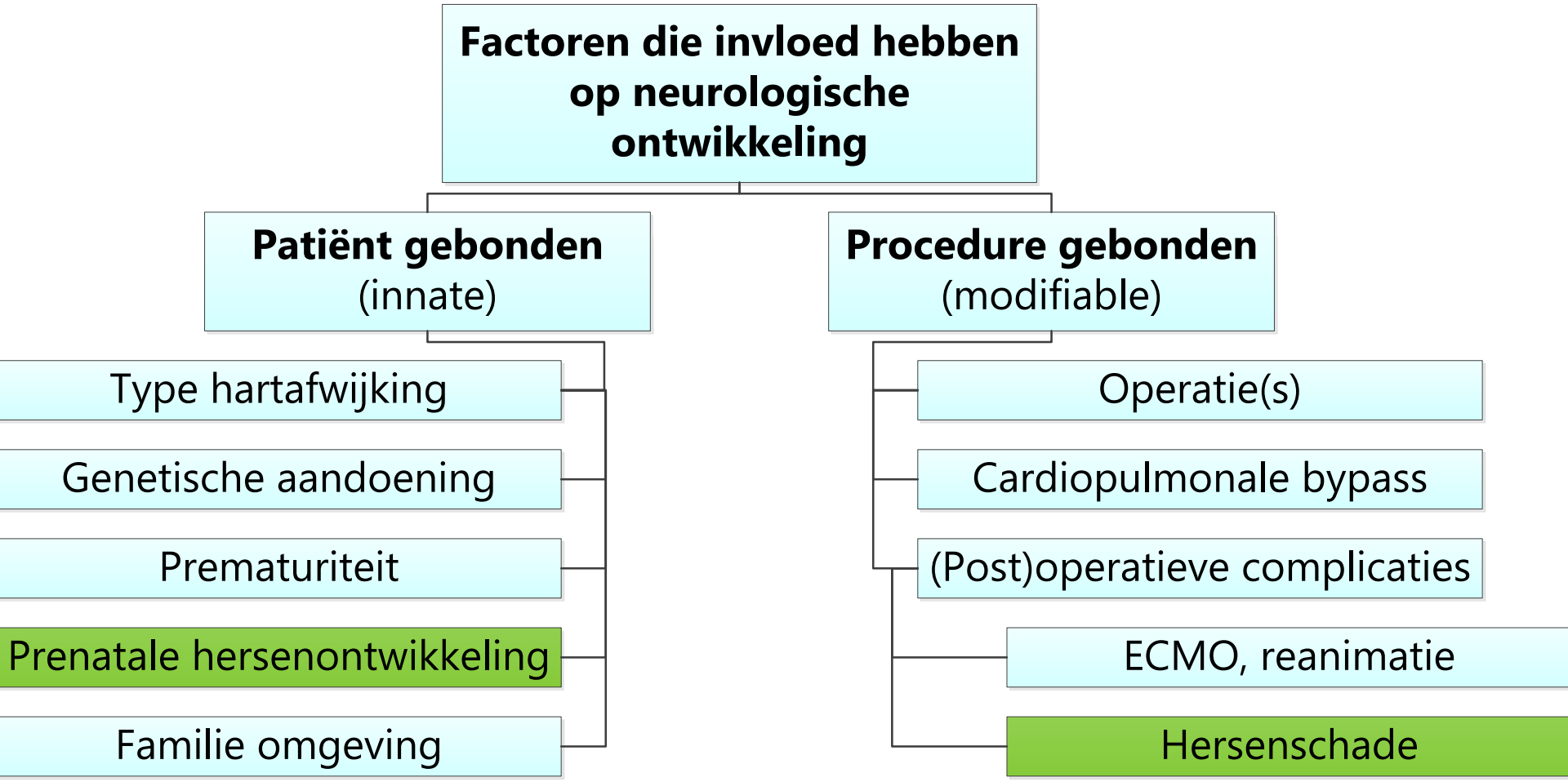
PREVENTING COLLATERAL DAMAGE

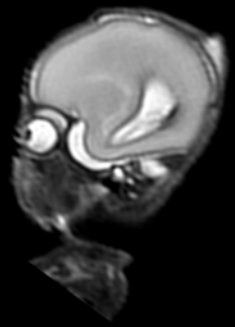


Modified from Hovels-Gurich, Front paediatr 2016

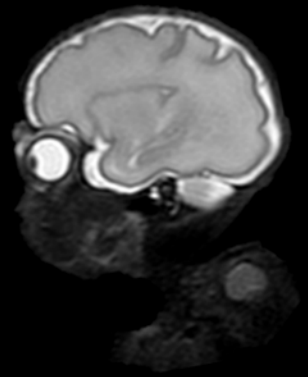


Modified from Hovels-Gurich, Front paediatr 2016

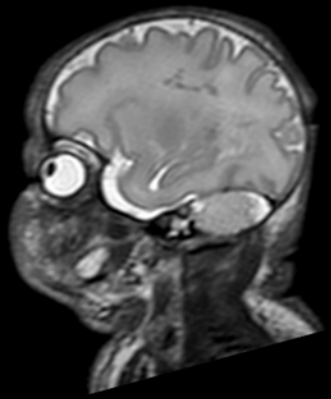
Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE



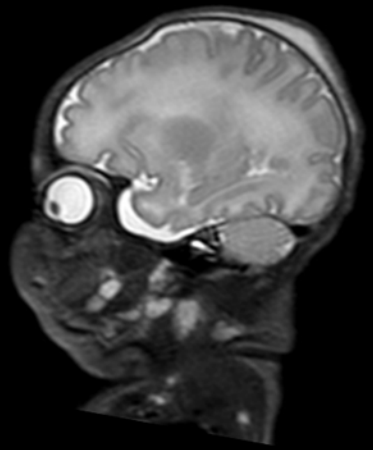
25 weken



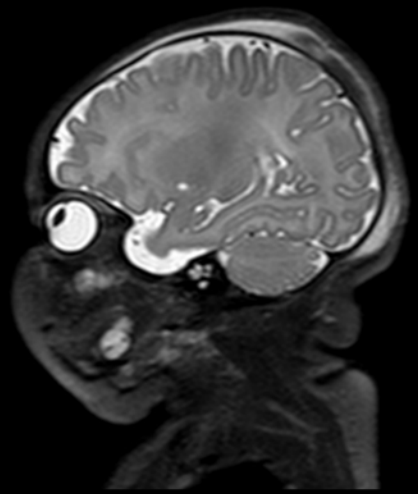
30 weken



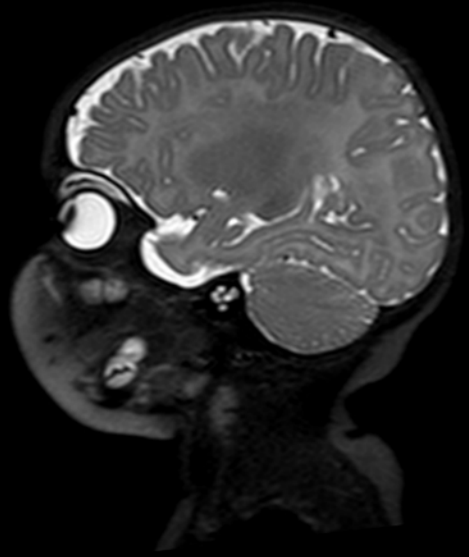
33 weken



36 weken

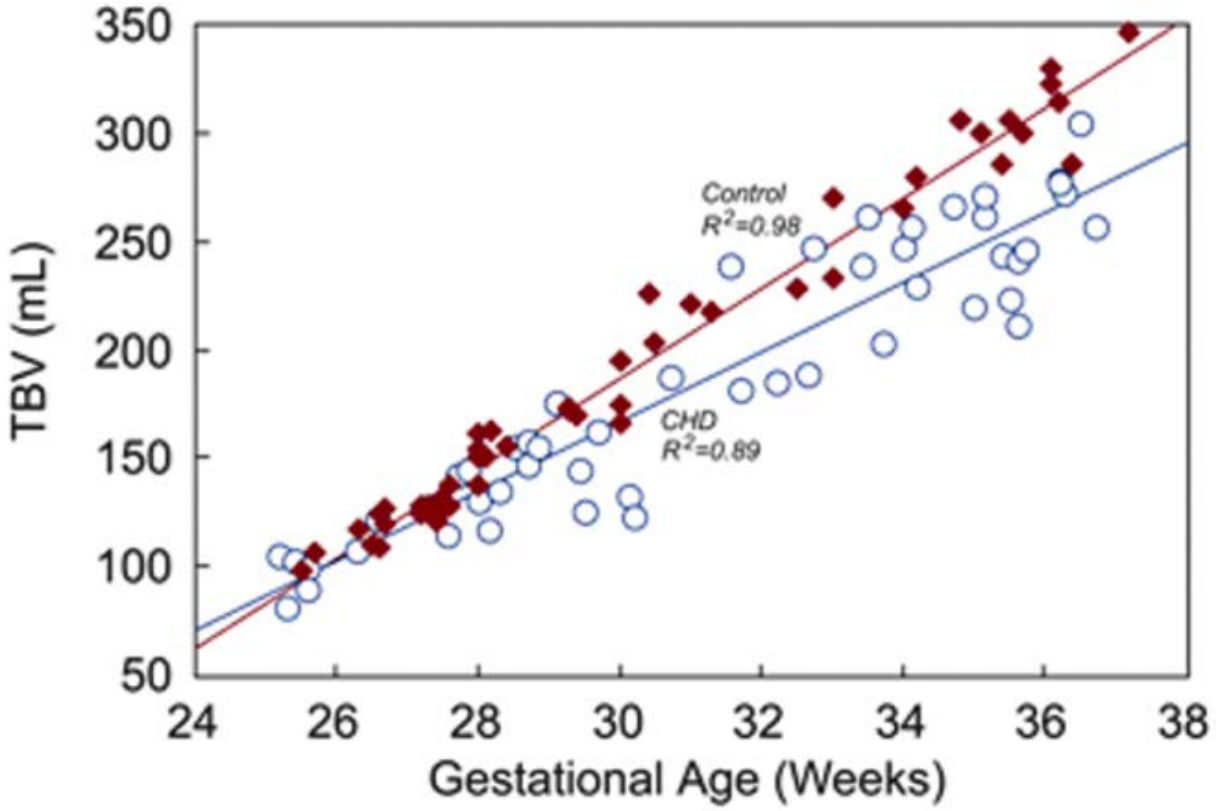


Uitgerekende datum



3 maanden

Foetale hersenontwikkeling



Limperopoulos, Circulation 2010.

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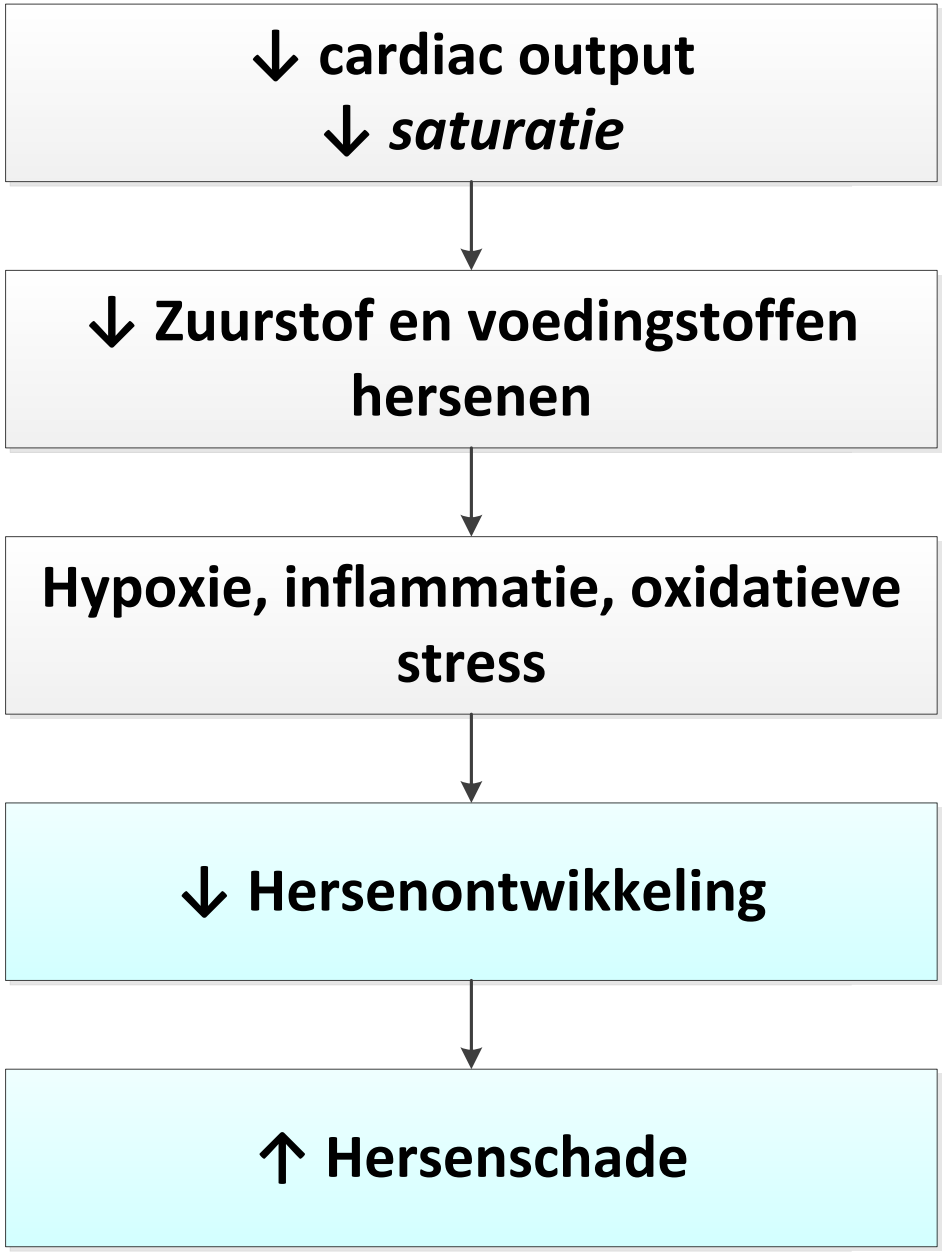
PREVENTING COLLATERAL DAMAGE



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PREVENTING COLLATERAL DAMAGE

Congenital Heart Disease Life Span

PREVENTING COLLATERAL DAMAGE

Wilhelmina Kinderziekenhuis

Child health, science for life

Beeldvorming

Crucial trial

Follow-up

Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE

Beeldvorming hersenen

Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE

1. Hartafwijking waarvoor een operatie nodig is in de eerste levensweken

- Hypoplastisch linker/rechter hart (HLHS, HRHS, DILV, tricuspidalis atresie, ongebalanceerd AVSD)
- Transpositie van de grote vaten
- Afwijking aortaboog (HLHC, IAA, HAA, CoA)
- TAPVC
- *Tetralogie van Fallot*
- *Truncus arteriosus*

2. Beeldvorming hersenen

MRI

Hersenbewaking
(EEG +NIRS)

Congenital Heart
Disease Life Span
PREVENTING COLLATERAL DAMAGE

Foetale MRI

Postnatale MRI

Postop MRI

20 weken Diagnose

40 weken Geboorte

Neonatale OK

Follow Up

Congenital Heart Disease Life Span

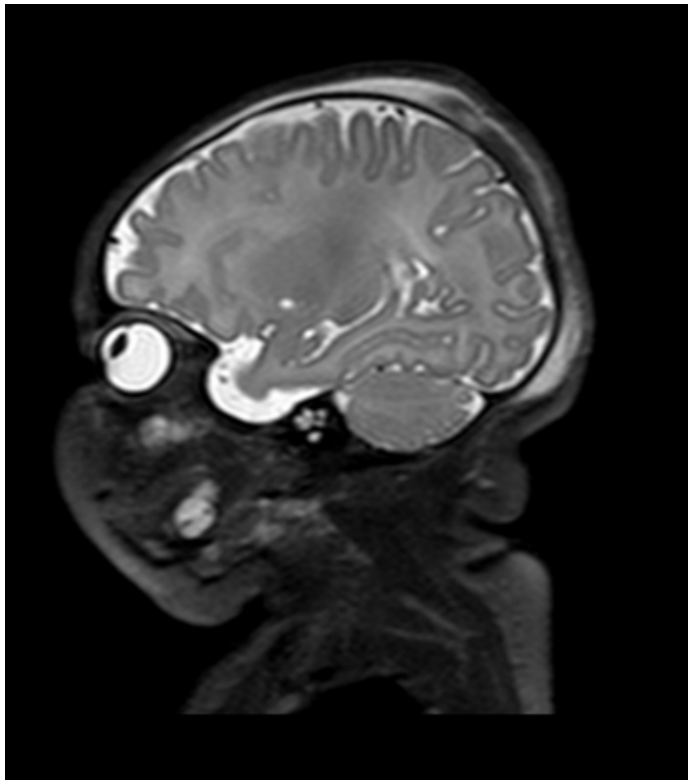
PREVENTING COLLATERAL DAMAGE

Foetale MRI:

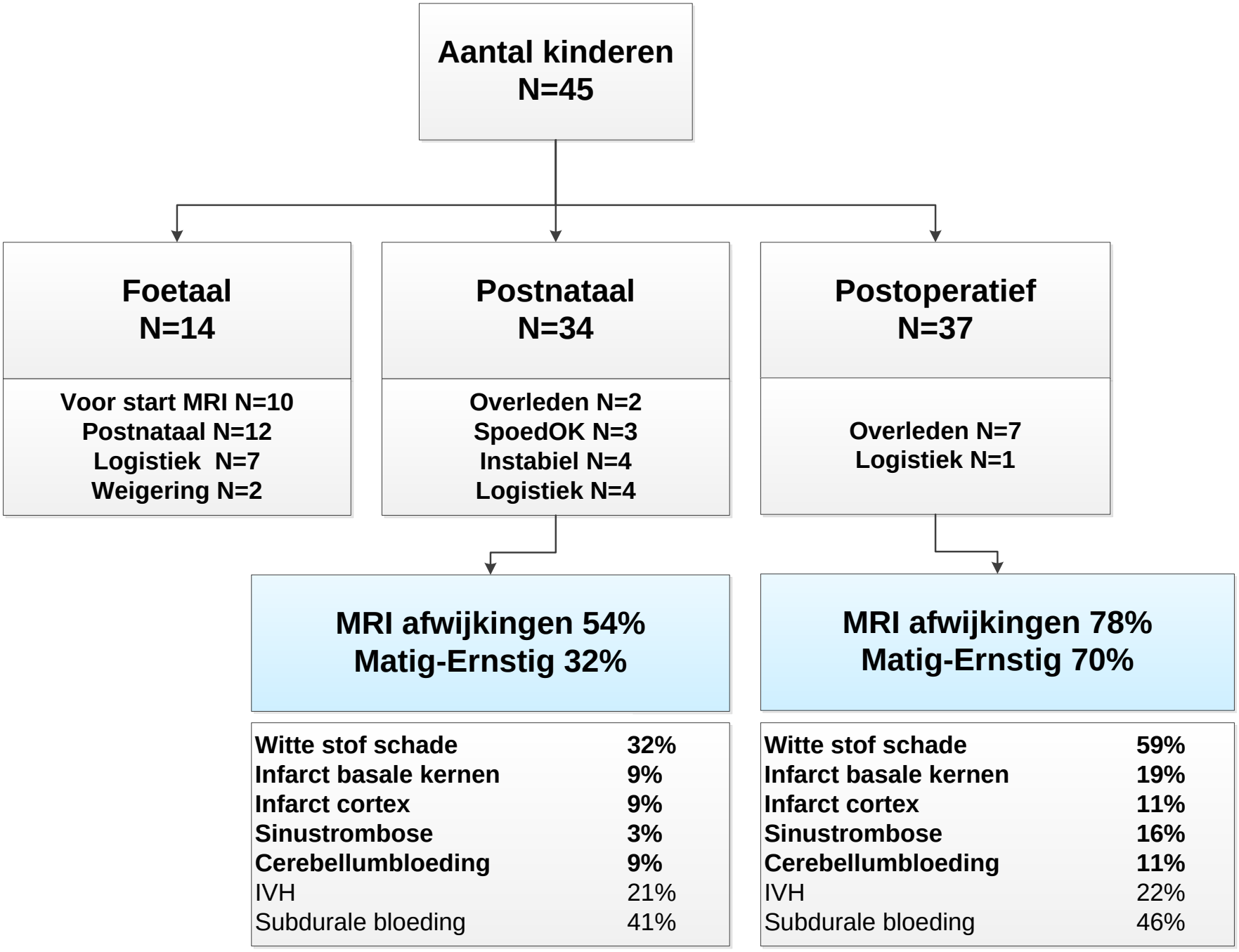
- Hersenontwikkeling?
- Aangeboren afwijkingen?

Neonatale MRI

- Hersenontwikkeling?
- Hersenschade?

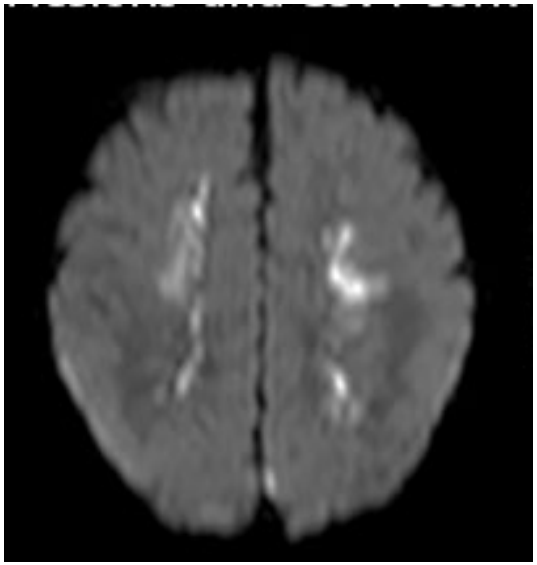


Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE


Aantal kinderen
N=45
Foetaal
N=14
Voor start MRI N=10
Postnataal N=12
Logistiek N=7
Weigering N=2
Postnataal
N=34
Overleden N=2
SpoedOK N=3
Instabiel N=4
Logistiek N=4
Postoperatief
N=37
Overleden N=7
Logistiek N=1
MRI afwijkingen 54%
Matig-Ernstig 32%
Witte stof schade 32%
Infarct basale kernen 9%
Infarct cortex 9%
Sinustrombose 3%
Cerebellumbloeding 9%
IVH 21%
Subdurale bloeding 41%
MRI afwijkingen 78%
Matig-Ernstig 70%
Witte stof schade 59%
Infarct basale kernen 19%
Infarct cortex 11%
Sinustrombose 16%
Cerebellumbloeding 11%
IVH 22%
Subdurale bloeding 46%


Radiologie
PICU

1



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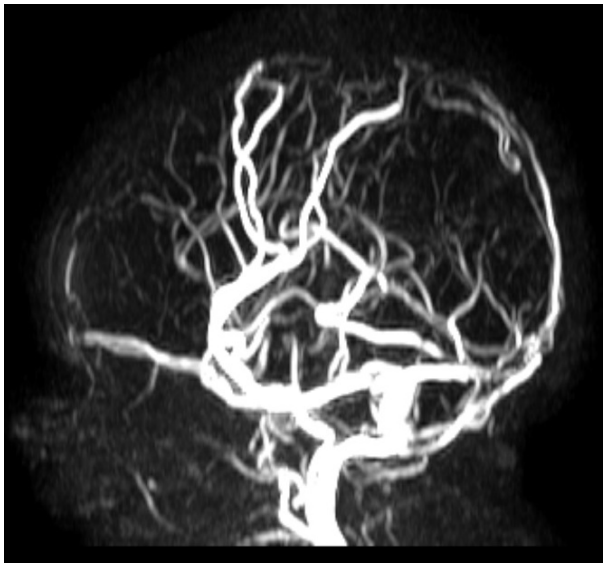

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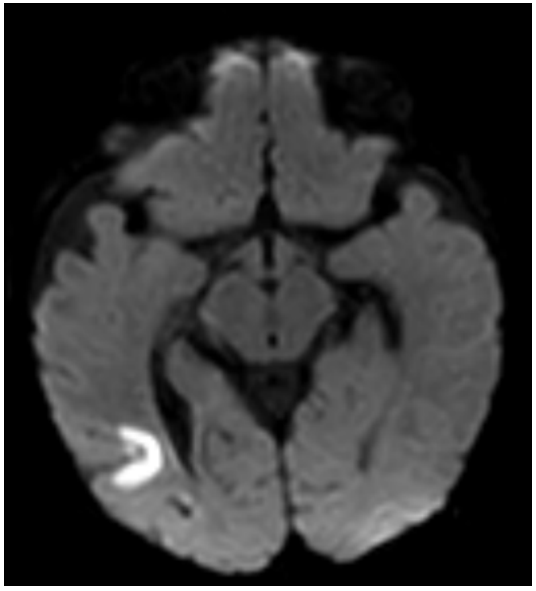

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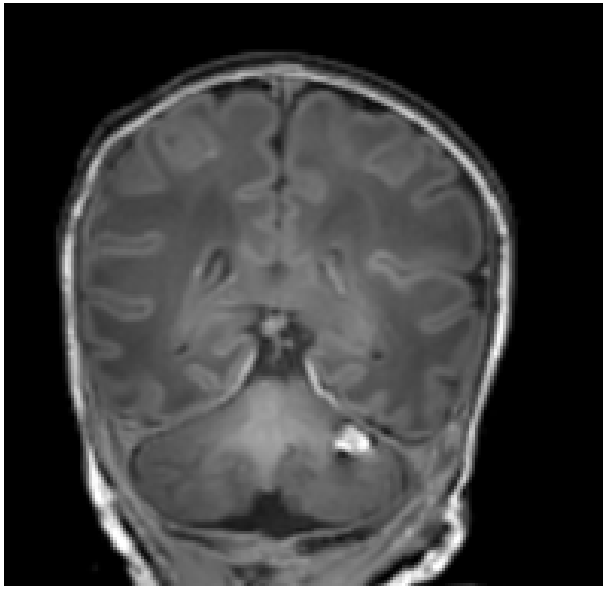

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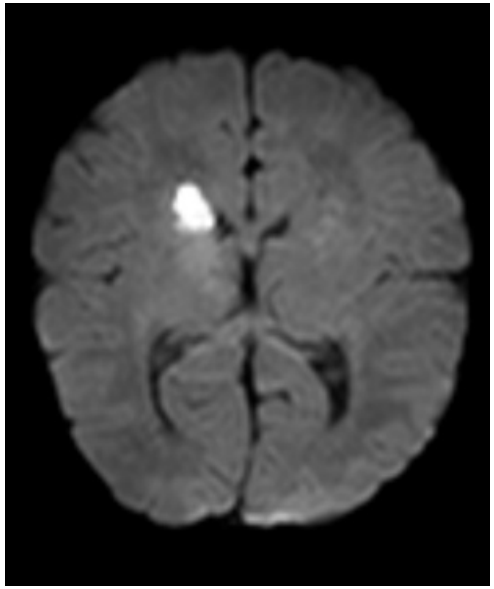

Congenital Heart Disease Life Span

PREVENTING COLLATERAL DAMAGE

Foetale MRI

Postnatale MRI

Postop MRI

20 weken Diagnose

40 weken Geboorte

Neonatale OK

Follow Up

EEG + NIRS

EEG + NIRS

Congenital Heart Disease Life Span

PREVENTING COLLATERAL DAMAGE

EEG: Electroencephalographie (hersenfilmpje)

> Functie van de hersenen

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PREVENTING COLLATERAL DAMAGE

NIRS: Near-infrared spectroscopy

> Regionale zuurstof saturatie hersenen

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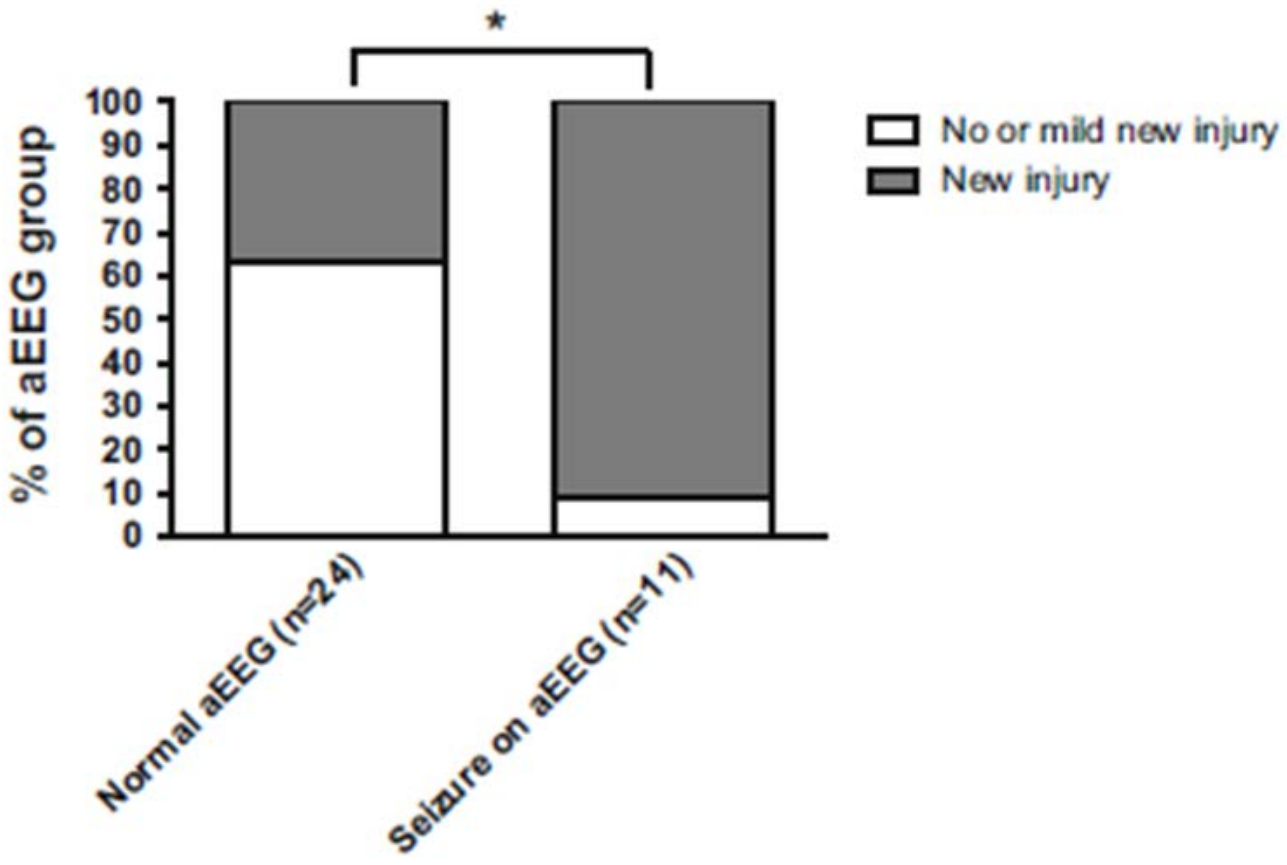
PREVENTING COLLATERAL DAMAGE

EEG onderzoek in neonaten met een congenitale hartafwijking



Algra, Intensive Care Med 2015

Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE

Beeldvorming

Crucial trial

Follow-up

Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE

Follow-up

Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE

Follow-up

- Uitgebreid motorisch en neurocognitief onderzoek bij alle mijlpalen
  - Voor school (2 jaar)
  - Voor groep 3 (5 jaar)
  - Voor middelbare school (11 jaar)
  - Einde middelbare school (18 jaar)

- Vergelijking met referentie cohort (YoUth Cohort)

- Interventies op jonge leeftijd

Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE

Follow-up

- **Relatie hersenschade op MRI – kliniek**
- Flexibel follow-up programma

Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE

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Congenital Heart Disease Life Span
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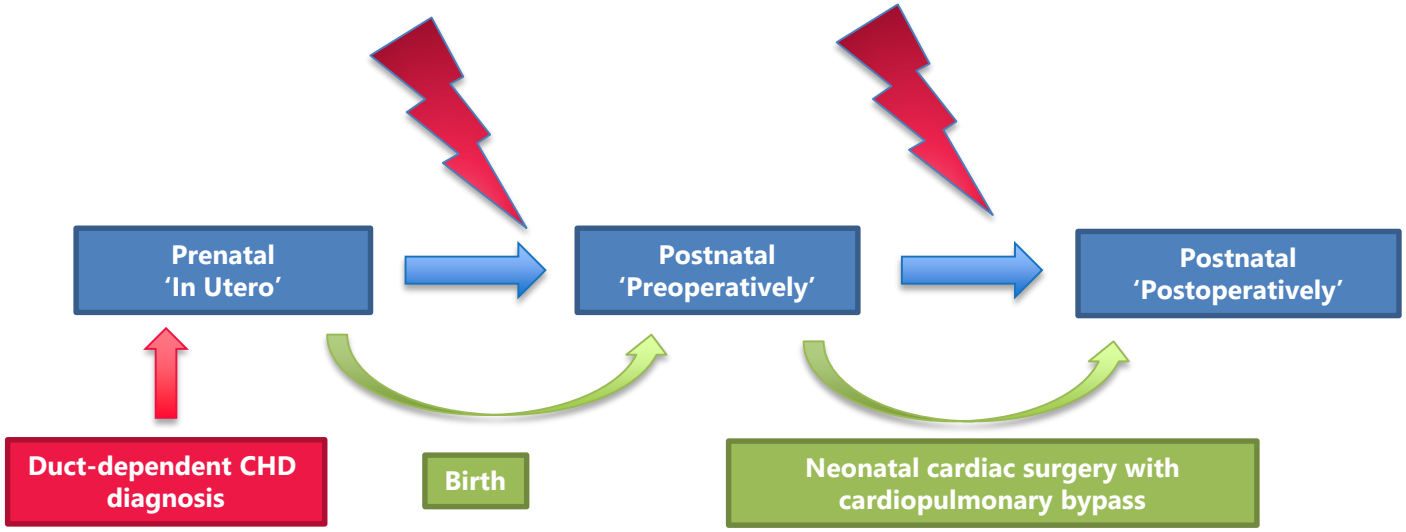
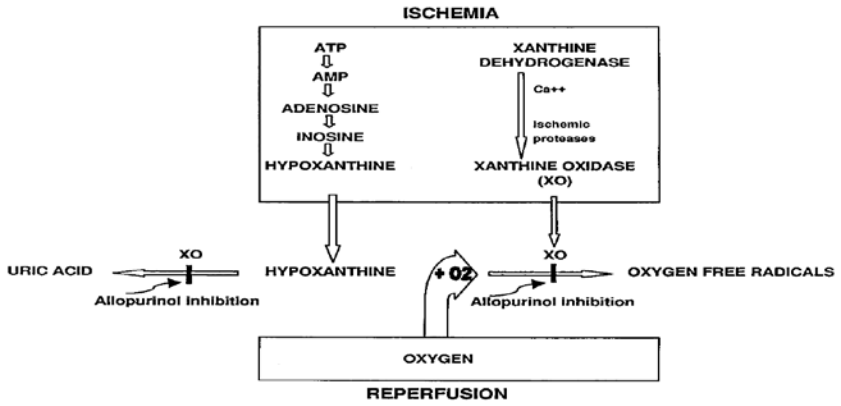


Introduction

- Allopurinol
  - Xanthine oxidase inhibitor
    - Prevents formation toxic oxygen free radicals
    - Scavenger of free radicals
    - Proradical chelator
  - Anti-inflammatory
  - Neuroprotective effects
  - Cardiovascular protective properties



Boda D. Pren Neon Med 1999

Van Bel F. Pediatrics 1998

Allison BJ. FASEB J 2016

Torrance HL. Pediatrics 2009

Benders MJNL. Arch Dis 2006

Clancy RR. Pediatrics 2001

Kaandorp JJ. Arch Dis 2005

Kaandorp JJ. Arch Dis 2012

Gunes T. Pediatr Neurol 2007

Derks JB. Pediatr Res 2010

Study design

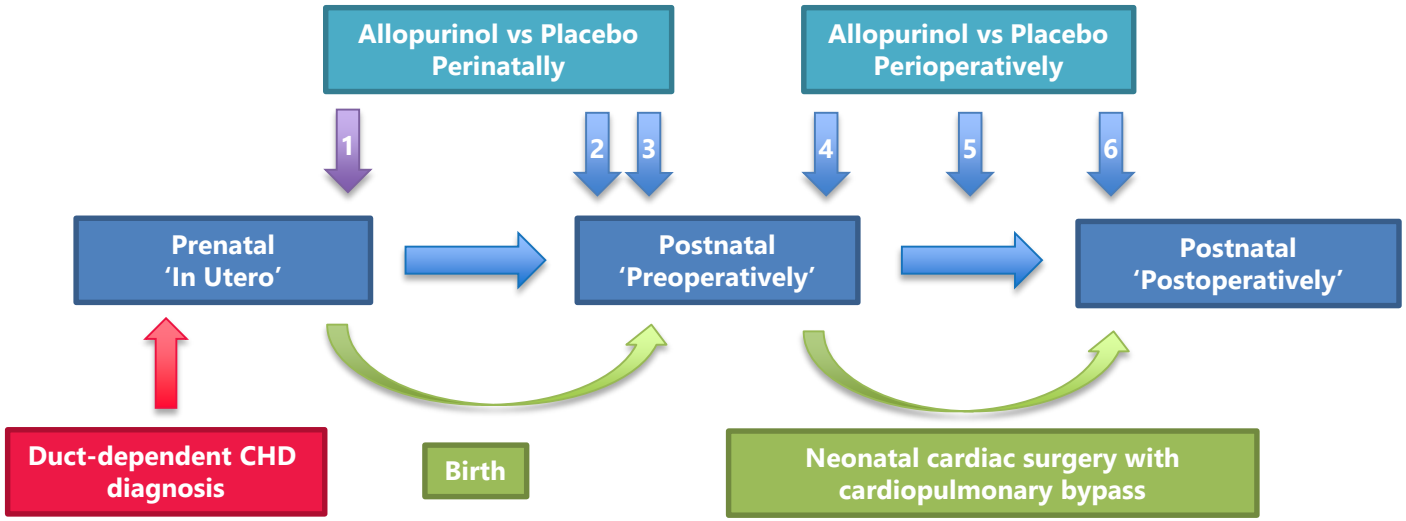
Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE
INTERVENTION

- Prospective, randomized, double blind, placebo-controlled, multicentre trial (phase III)
  - CAHAL Amsterdam/Leiden
  - EMC Rotterdam
  - UMC Groningen
  - UMC Utrecht

Congenital Heart Disease Life Span
PREVENTING COLLATERAL DAMAGE

Study medication

Antenatal diagnosis



INTERVENTION

Congenital Heart Disease Life Span

PREVENTING COLLATERAL DAMAGE

Conclusie

- Hersenschade komt frequent voor bij ernstige aangeboren hartafwijkingen
- Hersenschade geeft NIET altijd ontwikkelingsproblemen
- Ernstige hersenschade is op jonge leeftijd zichtbaar
- Minder ernstige hersenschade uit zich pas veel later en subtieler (complexe executieve functies)
- Gelukkig is het IQ van ouders meer voorspellend voor IQ van het kind dan de hartafwijking

Bedankt!

Wilhelmina Kinderziekenhuis

- Kinderhartcentrum
- Kinder-IC
- Neonatologie
- Obstetrie
- Radiologie
- Kinderbewegingscentrum
- Medische Psychologie

stichting hartekind