Appropriate Follow Up in Newborns with CHD

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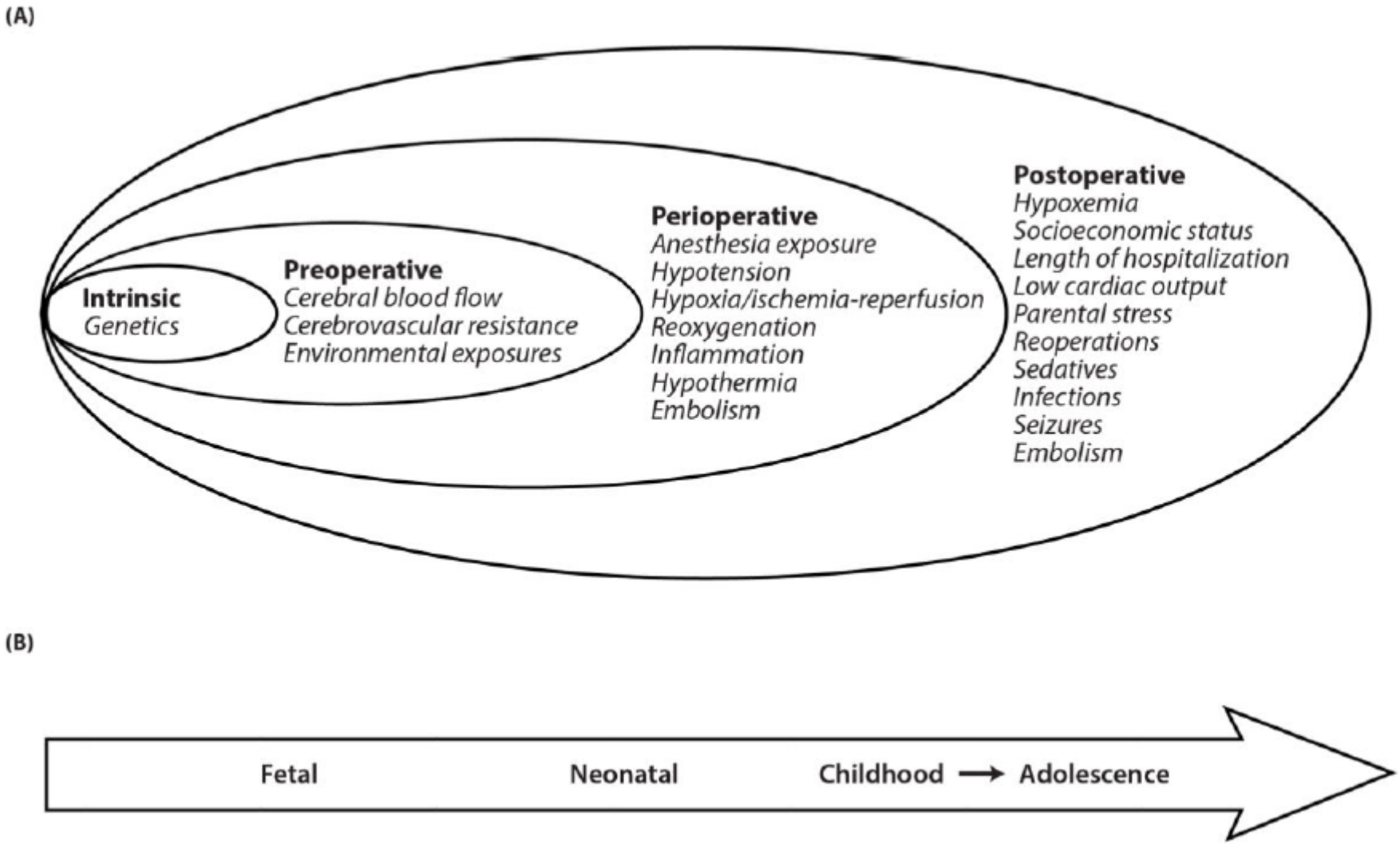
Neurodevelopmental Abnormalities and Congenital Heart Disease: Insights into Altered Brain Maturation



Morton et al, Circ Res 2017

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Introduction

- Incidence CHD: 6-8 /1000 live births (1% of live babies in the USA)
- Most frequent congenital malformation in childhood
- 1/3 CHD present genetic disorder (trisomy 21, 22q11microdeletion, CHARGE)
- ≅ 50% CHD require immediate surgical intervention
- 80%-90% survival rates even for complex CHD
- Good cardiac outcome→ more adults are alive now with CHD
- CHD are particularly vulneable for subsequent ***neurodevelopmental impairment ⟶ 50%***

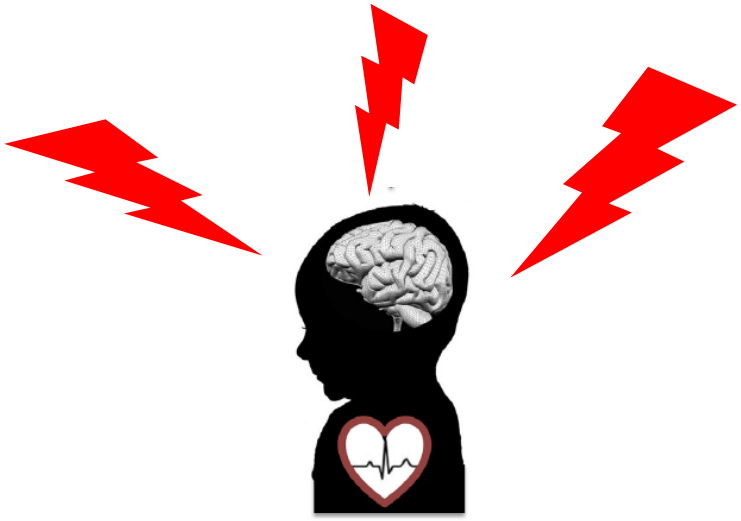
CHD and Brain Injury

The exact mechanism responsible for brain injury in CHD is not yet fully understood

Alterations in CBF and cerebral metabolism

Abnormalities in genetic pathways

Impaired cerebral oxygenation



Abnormal Cerebral Development

Miller et al. N Engl J Med 2007
MJ Mebius et al. Pediatric July 2017
B Latal. Clin Perinatol 43 (2016)
NHP Claessens et al. Neuroimage:Clinical 21 (2018)

CHD and Brain Injury

Reduction in total brain volume → Smaller head circumferences (at least 10% in HLHS/ToF)

White Matter → puntate lesions (PVL, stroke)

early microstructural abnormalities

Grey Matter → degree of cortical folding (gyrification)

Basal ganglia and thalami are smaller (TGA/SVP)

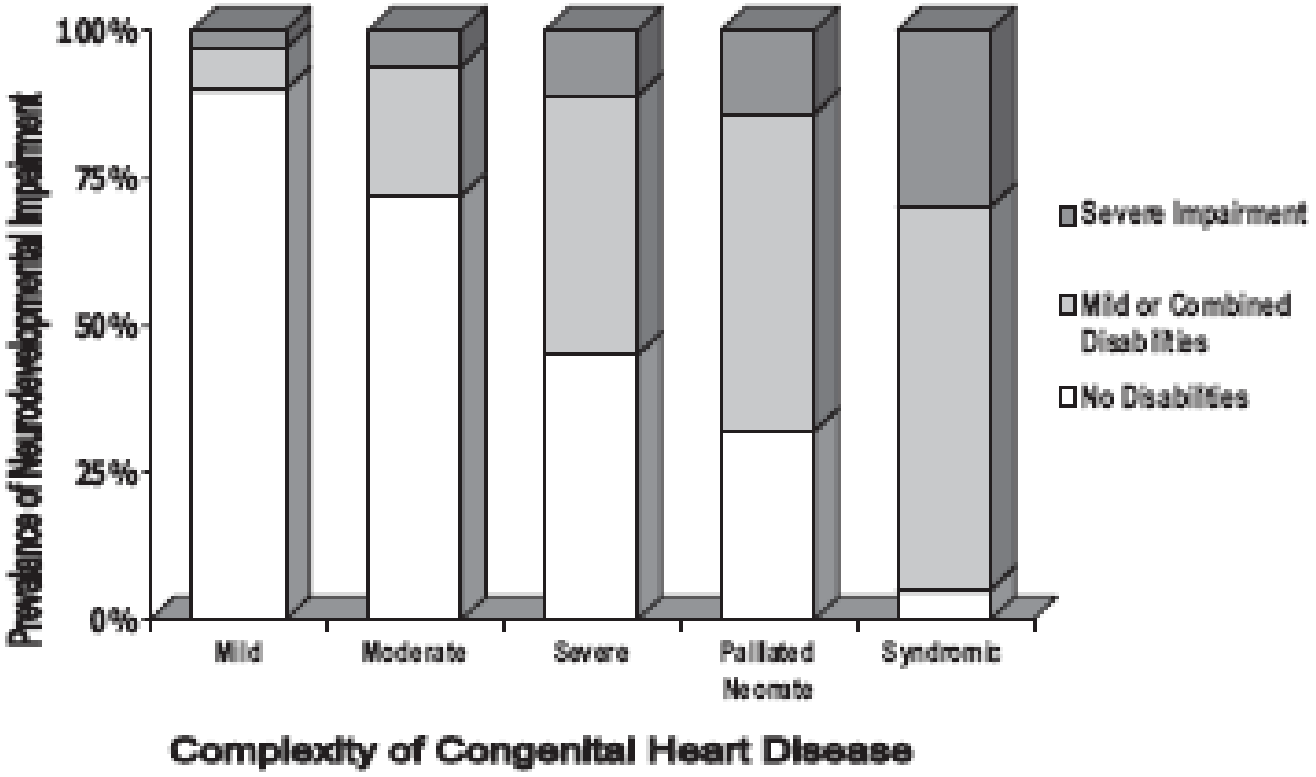
Reduction of 20% cerebellar volume (SVP/TGA)

S Pevandi et al. Neuroimage 185 (2019)
NHP Claessens et al. Neuroimage:Clinical 21 (2018)
MJ Mebius et al. Pediatric July 2017

Neurodevelopmental Outcomes in Children With Congenital Heart Disease: Evaluation and Management

A Scientific Statement From the American Heart Association

This statement has been approved by the American Academy of Pediatrics.



BS Marino at al. DOI: 10.1161/CIR.0b013e318265ee8a

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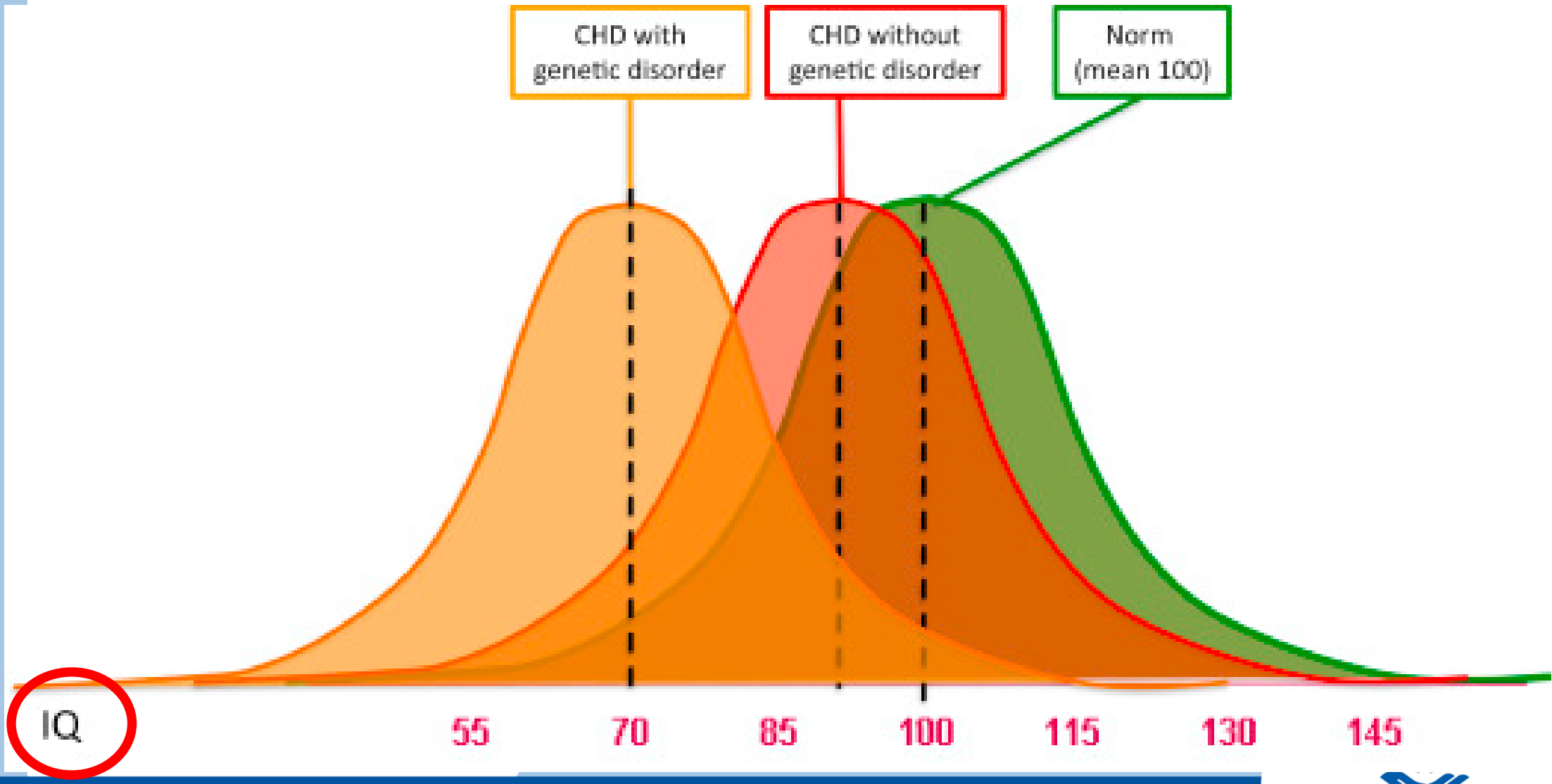
Neurodevelopmental Outcomes of the Child with Congenital Heart Disease



B Latal. Clin Perinatol 43 (2016)

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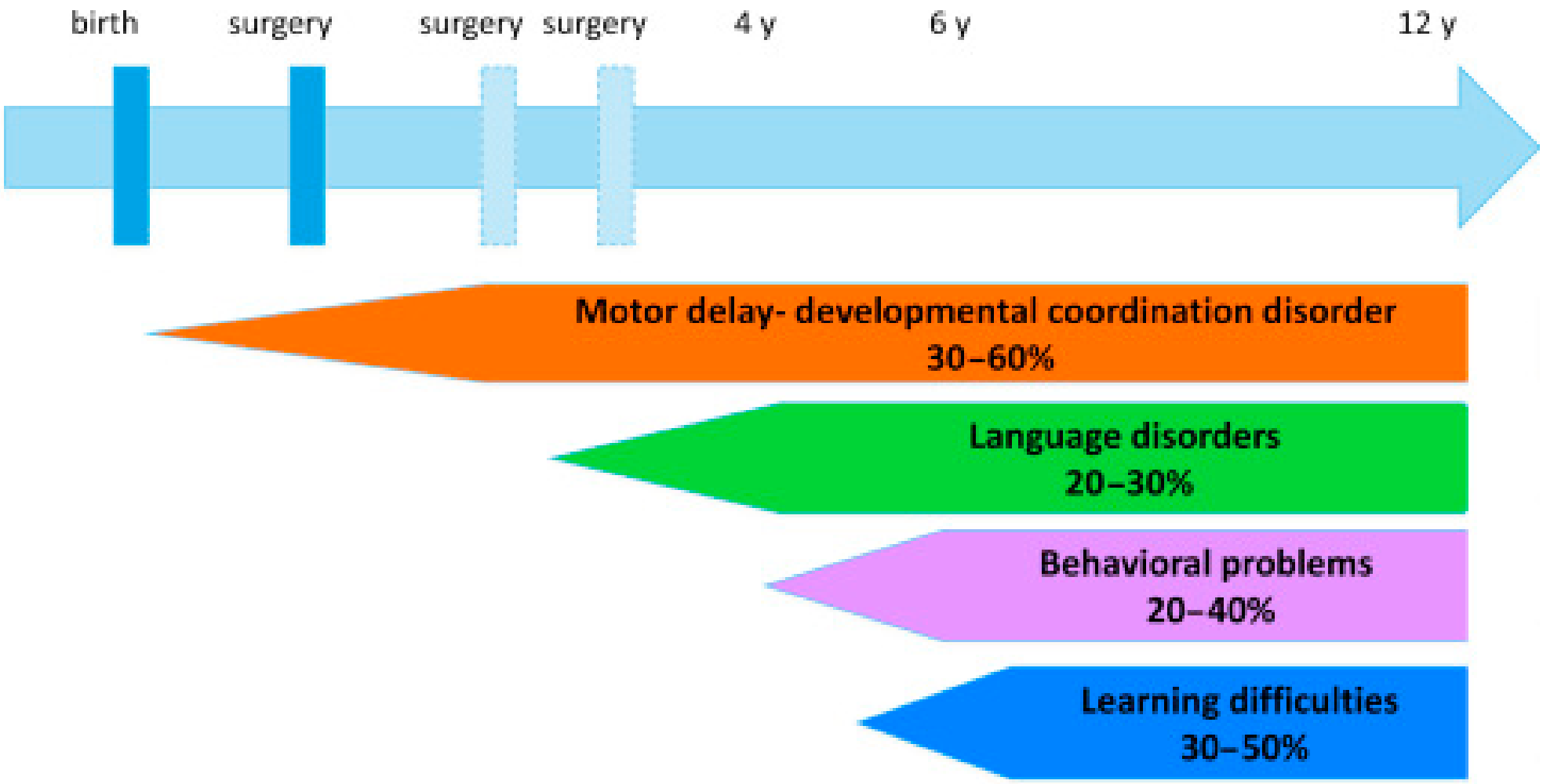
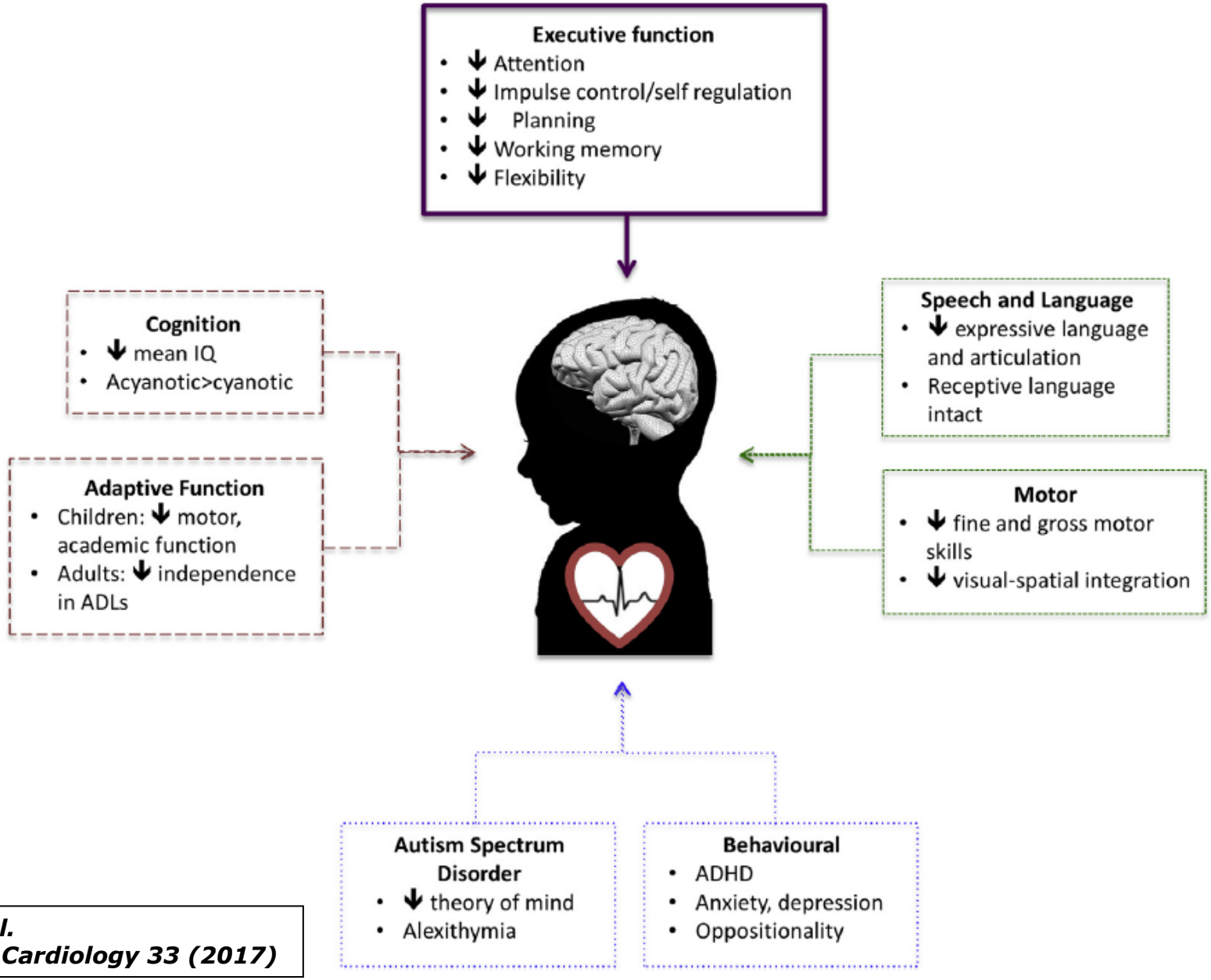
Neurodevelopmental Outcomes of the Child with Congenital Heart Disease



B Latal. Clin Perinatol 43 (2016)

Congenital Heart Disease and Neurodevelopment: Clinical Manifestations, Genetics, Mechanisms, and Implications



SN Nattel et al.
Canadian J of Cardiology 33 (2017)

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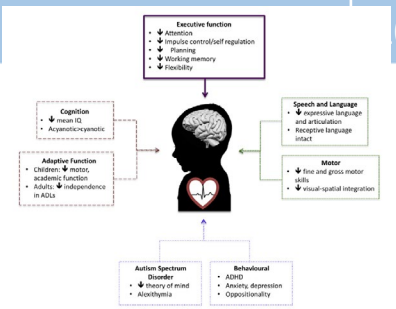
Neurodevelopmental outcome



Lower IQ (>10 points in cyanotic CHD)

Motor deficits 20-50%(cyanotic and acyanotic CHD):

- **HLHS -2SD at 1 year**
- **fine and gross motor skills at 18 months and 5 years**

Deficits visual-spatial/visual-motor integration (TGA school-age)

Language: speech delay of 2-4 months

- **apraxia (ToF)**
- **poor language (cyanotic CHD)**

Behavioural comorbidities:

- **autism spectrum disorder**
- **ADHD (cyanotic CHD)**
- **somatization and aggressive behaviour**
- **depression/anxiety**

SN Nattel et al. Canadian J of Cardiology 33 (201

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Appropriate Follow Up in CHD

- Cardiological assessment
- Nutritional assessment
- Respiratory assessment
- *Neurocognitive and neuromotor outcome*

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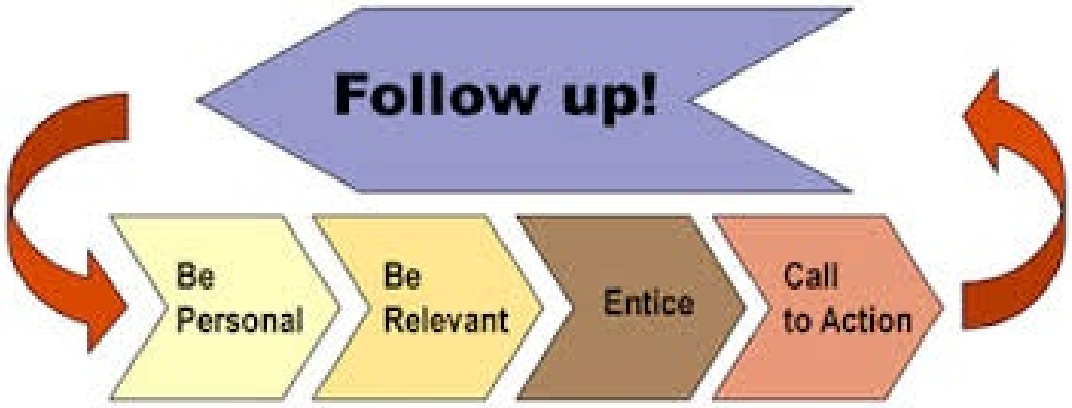
Appropriate Follow up in CHD

In specialized and expert centers



Multidisciplinary

- **Developmental Neonatologist/Pediatrician**
- **Cardiologist**
- **Pediatric Nurse**
- **Neurologist**
- **Neuropsycologist**
- **Physiotherapist and Speech Therapist**

Neurodevelopmental Outcomes of the Child with Congenital Heart Disease

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.

B Latal. Clin Perinatol 43 (2016)

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CHD FU in OPBG
From 2004 DNMC

1. Dedicated
2. Multidisciplinary
3. *Stable team* →continuity of care
4. Counseling and care for child and parents
5. Life long follow up
6. Research projects and clinical-scientific collaborations

Post discharge outpatient clinic → TO → Day Hostital

CHD Follow Up OPBG

NEONATOLOGIST

CARDIOLOGIST

GASTROENTEROLOGIST
PNEUMOLOGIST
GENETICIAN
RADIOLOGIST
NEUROLOGIST
NEUROPSYCOLOGIST
PHYSIOTHERAPIST

FU Time Points OPBG

1. One week after discharge from hospital
2. Three months
3. Six months
4. Nine months (if necessary)
5. Twelve months

DNMC

>1 YEAR → CARDIOLOGIST FU PROGRAM DMCCP

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Post Discharge Visit

✓ **Feeding**

- Water and caloric intake
- device: PEG, NGT
- Gastroenterological and auxological assessment (statural/weight growth)

✓ **Therapy**
✓ **Vaccinations/ RSV prophylaxis**

✓ **Family care**
✓ **Check of the contacts with attending pediatrician**

✓ **cardiological and cardiac surgery program**

✓ **Follow up program**

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CHD FU DNMC OPBG

OUTCOME	3 mos	6 mos	9 mos	12 mos	18 mos Outpatient clinic DNMC	24 mos Outpatient clinic DNMC	4→12 yrs Outpatient clinic DNMC
Auxological	+ Blood sampling	+ Boold sampling	+	+	+	+	+
Respiratory	PFR	PFR		PFR			
Cardiac	+	+	+	+	+ *DH DMCCP*	+ *DH DMCCP*	+ *DH DMCCP*
Genetic	+			+			
Ultrasound	+	+	+	+			
Visual				+			
Audiological		+		+			
Neuromotor Hammersmith exam	+	+	+	+	+	+	Neurologist
Physiotherapist	+	+		+	+ *DH DMCCP*	+ *DH DMCCP*	+ *DH DMCCP*
Neurodevelopmental		+ BAYLEY III	+	+ BAYLEY III	+ BAYLEY III	+ BAYLEY III	+ *DH DMCCP*
Family care	+	+	+	+	+	+	+
Neuroimmaging				+			

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OUTCOME CHD FU DNMC
Bayley III at 12 mos

FU AT 12 MOS (n)	80/2yrs
EG (w)	37,7 (33-40)
PN (g)	2839 (900-3850)
DP (%)	81% (65/80)
Genetic abnormalities (n)	3
Stay at hospital (dd)	33 (7-120)
Surgery (n)	2 (1-3)
Cognitive	98,2
Speech (RC+EC)	97,3
RC	9,42
EC	9,76
Motor	85,7
Fine motor	8,86
Gross motor	6,86

PRENATAL COUNSELING

Family care

Child

Establish a good ralationship with the parents

Provide adeguate information on the desease and the clinical and surgery course

Psycologist → Evaluation of the state of anxiety

Provide an optimal managment during pregnancy and delivery

Prenatal diagnosis → earlier use of PGE → less microstructural abnormalities of WM
(*Peyvandi et al. JAMA Pediatr 2016)*

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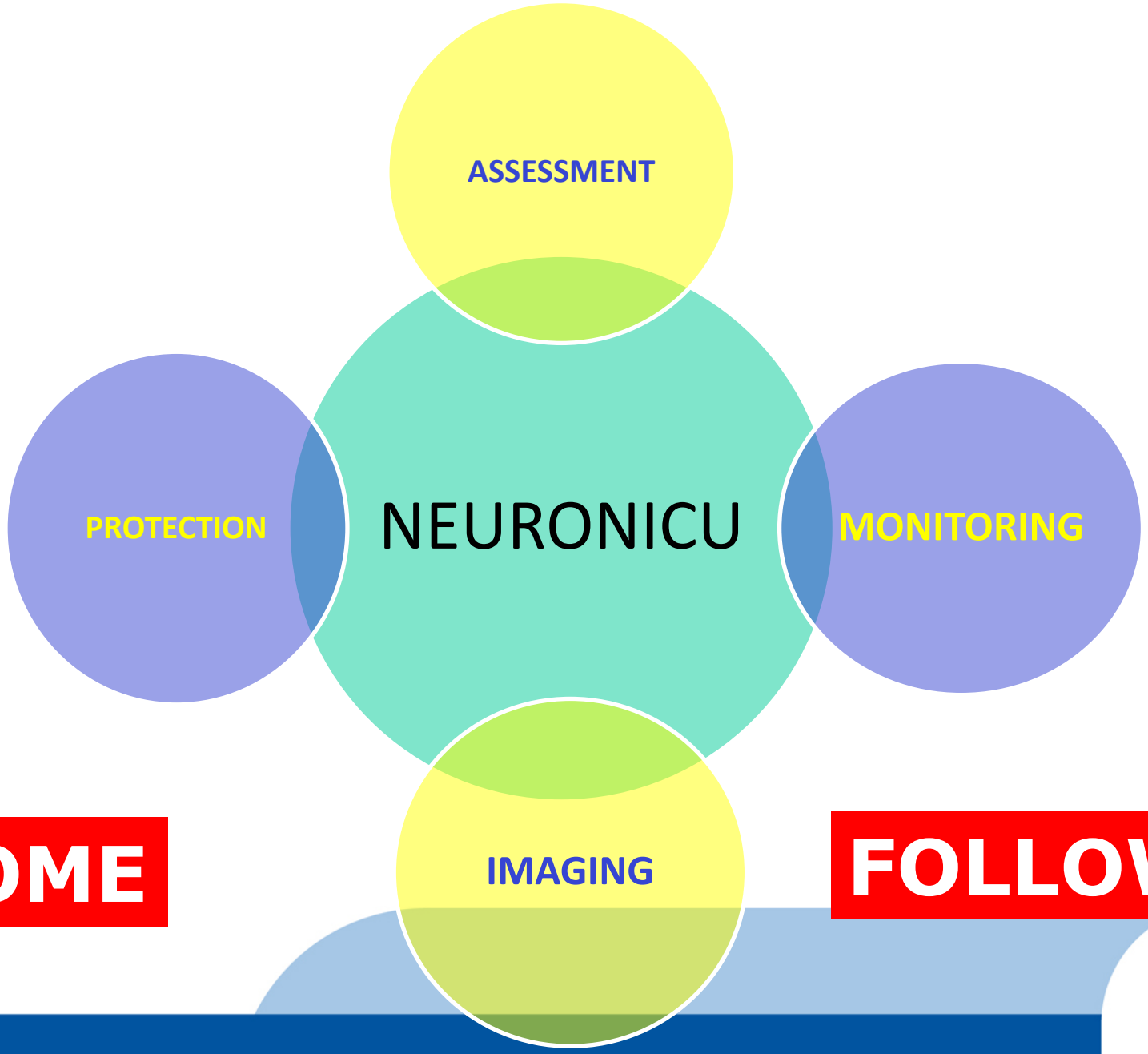
NEURONICU 4 DOMAINS



OUTCOME

FOLLOW UP

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Development of a NeuroNICU with a Broader Focus on All Newborns at Risk of Brain Injury: The First 2 Years

Krisa P. Van Meurs, MD[1] Elisabeth S. Yan, BS[1] Kathi S. Randall, NNP, RN, CNS[2]
Valerie Y. Chock, MD, MS[1] Alexis S. Davis, MD, MS[1] Cecelia S. Glennon, RN, NNP[2]
Catherine L. Clark, PhD[1,3] Courtney J. Wusthoff, MD, MS[3] Sonia L. Bonifacio, MD[1]

Am J Perinatol

The mortality rate was 11% for our NeuroNICU population compared with 6% for the general NICU population during the same time period. The mortality rate was twofold higher in the subset of patients with a primary medical concern as compared with that of patients with a primary neurologic diagnosis (14 vs. 5%; $p < 0.01$). Deaths in the NeuroNICU were due to cyanotic congenital heart defects (37%), extreme prematurity (23%), and HIE (7%). The mortality rate for moderate to severe HIE was 8%.

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Development of a NeuroNICU with a Broader Focus on All Newborns at Risk of Brain Injury: The First 2 Years

Primary diagnosis	*n* (%)
Neurologic	205 (37.5)
Seizure	57 (10.4)
HIE/cooling	50 (9.2)
Neurologic abnormalities without specific diagnosis	40 (7.3)
CNS anomalies	24 (4.4)
Grade III or IV IVH or hydrocephalus	13 (2.4)
Meningitis	11 (2)
Neural tube defects	8 (1.5)
Stroke	2 (0.4)
Medical	341 (62.5)
Cyanotic CHD	162 (29.7)
Prematurity, gestational age ≤ 28 wk	100 (18.3)
Cardiorespiratory instability	55 (10.1)
Metabolic disease	13 (2.4)
ECMO/pre-ECMO	11 (2)
Total	546 (100)

Diagnosis	Monitoring	Neurology consult
HIE/cooling	aEEG, cEEG, and NIRS	Yes
Seizures	aEEG and cEEG	Yes
ECMO/pre-ECMO	NIRS and consider aEEG	As needed
Grade III/IV or hydrocephalus	aEEG	Yes
Critical/unstable	NIRS and consider aEEG	As needed
Preemie ≤ 28 wk	NIRS	As needed
CNS anomalies	cEEG and/or aEEG	Yes
Metabolic disease	cEEG and/or aEEG	As needed
Cyanotic CHD	NIRS	As needed
CNS infection	cEEG and/or aEEG	Yes
Symptomatic PDA	NIRS	As needed
ALTE/BRUE	aEEG	As needed
Hyperbilirubinemia > 20 or hemolytic process	NIRS and consider aEEG	As needed

FU for CHD at Development Risk

Neonatologist

- Auriti Cinzia
- Braguglia Annabella
- Calzolari Flaminia
- Campi Francesca
- ***De Marchis Chiara***
- Monaco Francesca
- Piersigilli Fiammetta
- Rechichi Jole
- Savarese Imma
- Savignoni Ferdinando

Psycologist

- Aite Lucia
- Bevilacqua Francesca
- ***Bucci Silvia***
- Coletti Maria Franca

Nurse

- Santi Roberta
- Stabile Tina
- Nicastro Marina
- Fierro Stefania
- D'Astore Michela

Pediatric Cardiologist

- Luciano Pasquini
- Alessandra Toscano
- Marco Campanale
- Alessia De Nisco

Pulmonary Function Test

- Calzolari Flaminia
- Columbo Claudia
- Landolfo Francesca
- Savignoni Ferdinando

DNMC - OPBG

Cardiologist Department

Care Givers

Pediatrians of National Health System

Family Association

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TAKE HOME MESSAGES

- Neonates and infants with CHD are at substantial risk of negative neurodevelopmental and neurobehavioral outcome → *dedicated and specific FU is mandatory*
- FU should be provided *untill school-age* and *ideally* untill *adolescence*
- *Family care is very important* in order to support parents from prenatal counseling to post discharge FU
- Strict neonatal monitoring of the neurological state of the brain is indicated pre and post surgery period (*cerebral MRI, NeuroNICU*)
- "…Information on timing of brain dysmaturation and acquired brain injury in CHD can be used in the development of strategies to optimize neurodevelopment…allopurinol, erythropoietin,melatonin…and may reduce damage induced by reoxygenation and reperfusion in the brain"

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