? 

predict.

prevent.

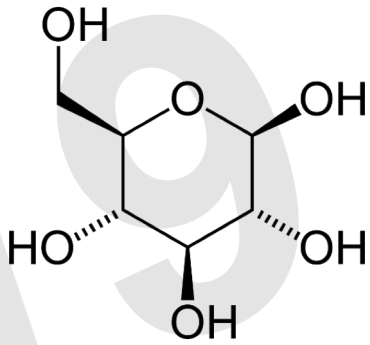
Endocrine emergencies in children

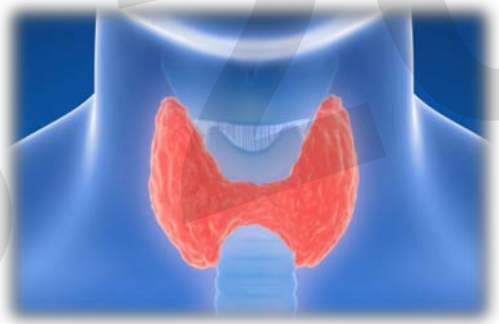
Philippe Lysy MD PhD MSc

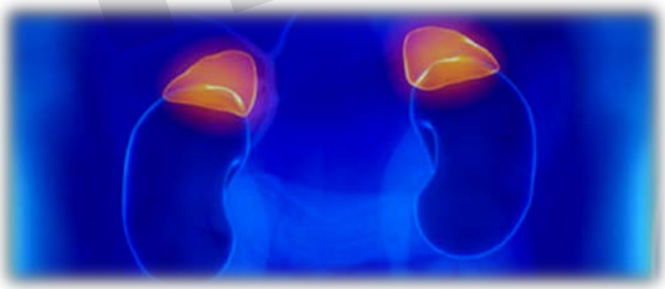
Laboratoire de pédiatrie, IREC

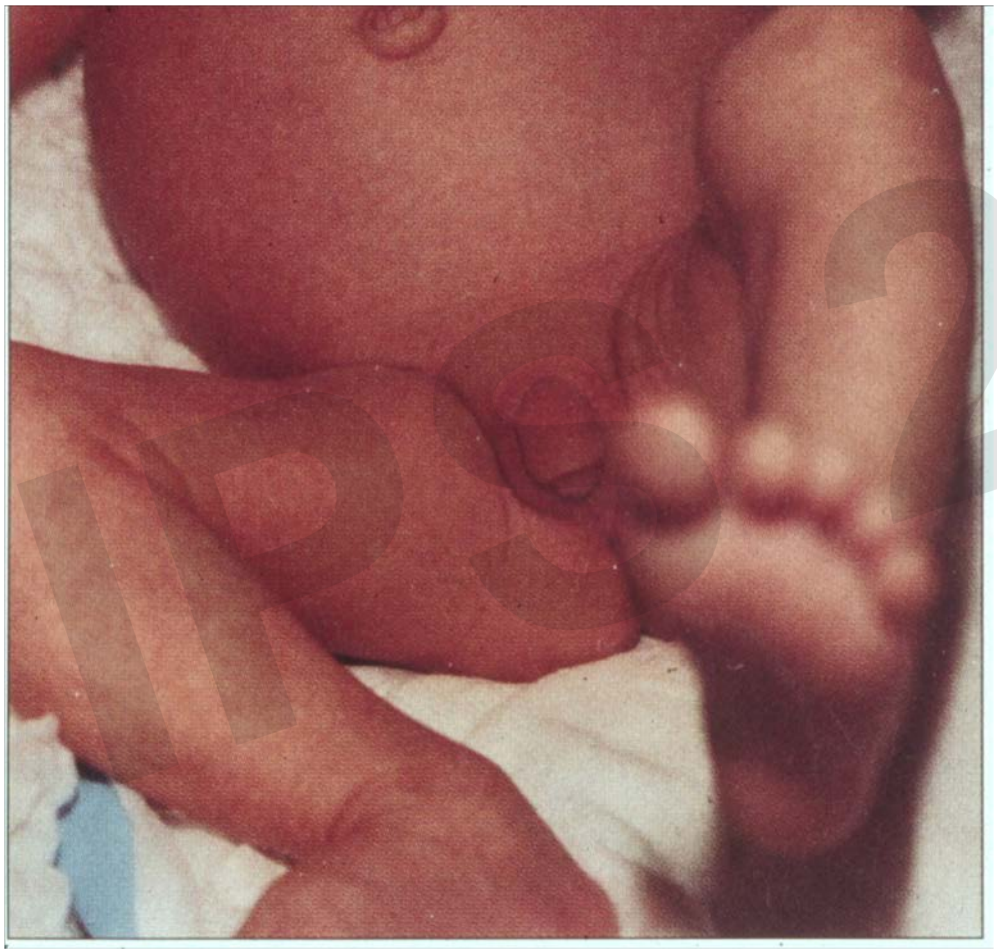
Unité d'Endocrinologie Pédiatrique

Endocrine Emergencies









10 yo boy

Hypospadias

Undescended testis

Na^+ 130 mM

K^+ 6.5 mM

Bicar 8 mM

What is your diagnosis?

The issues:

Incidence fails to decrease (4.4 - 17/100 patient-years)

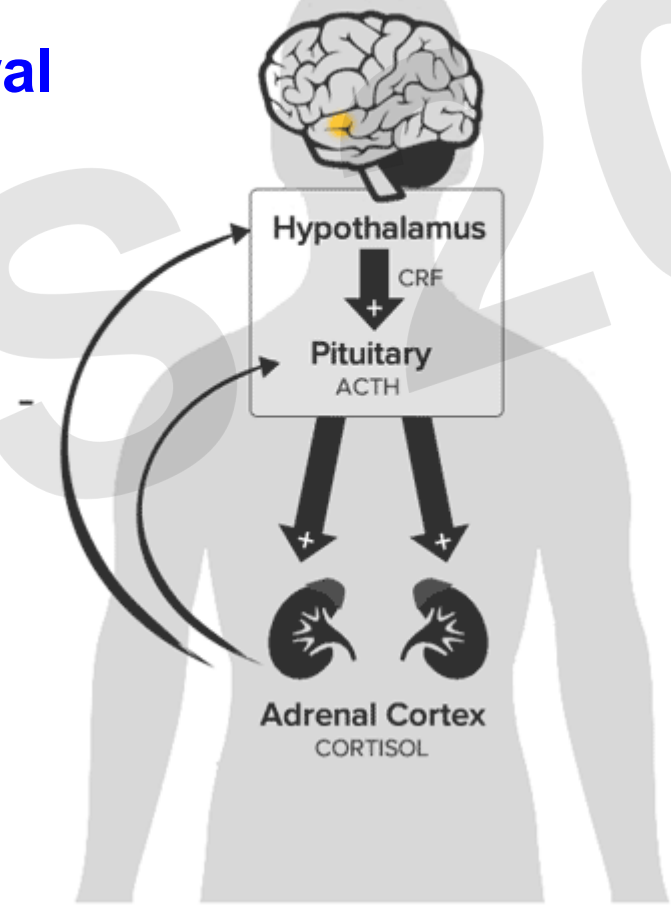
Death of 1 in 200 patients per year

Precipitating factors=
gastrointestinal disease (33%), infection (24%), surgery

Secondary insufficiency

Congenital, tumor, autoimmune,

GC R/-withdrawal



Primary insufficiency

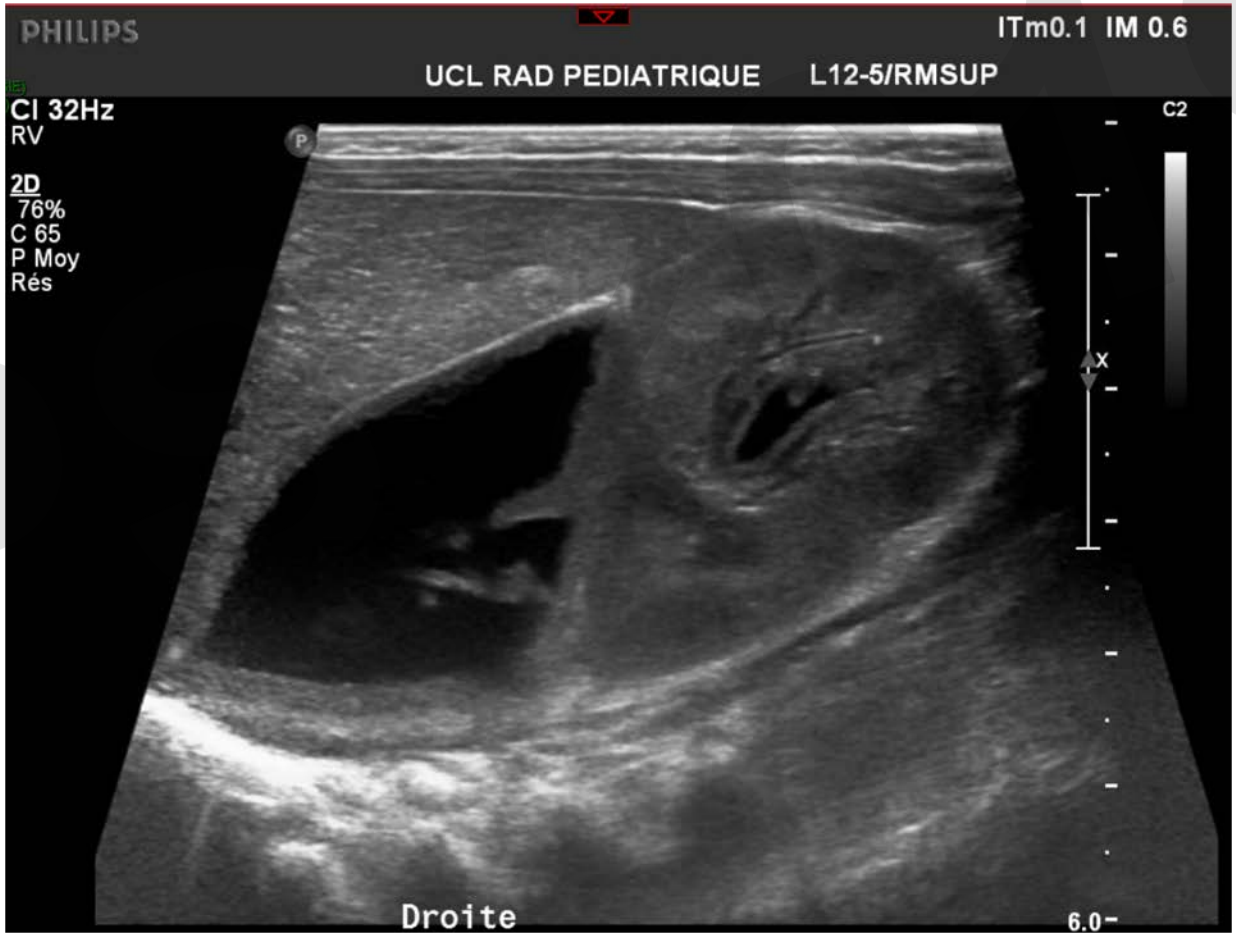
GC R/-withdrawal

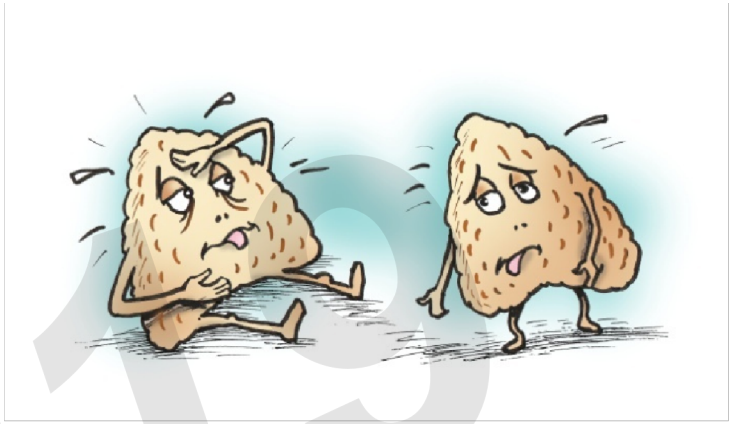
Autoimmune

Infections

Hemorrhage

Congenital (hypoplasia, adrenogenital syndrome, adrenoleucodystrophy)


PHILIPS
ITm0.1 IM 0.6
UCL RAD PEDIATRIQUE L12-5/RMSUP
CI 32Hz
RV
2D
76%
C 65
P Moy
Rés
C2
Droite
6.0



Symptoms/signs

Weakness, fatigue, lethargy

Abdominal pain (86%)

Vomiting (diarrhea)

Anorexia

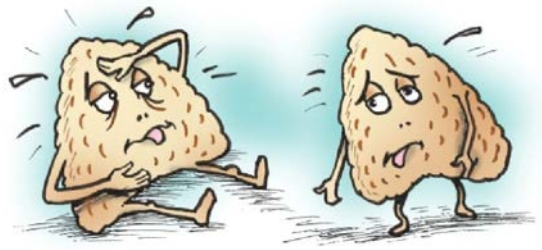
Fever (66%)

Pigmentation

Hypotension (>90%)

Shock

Muscle pain



Lab

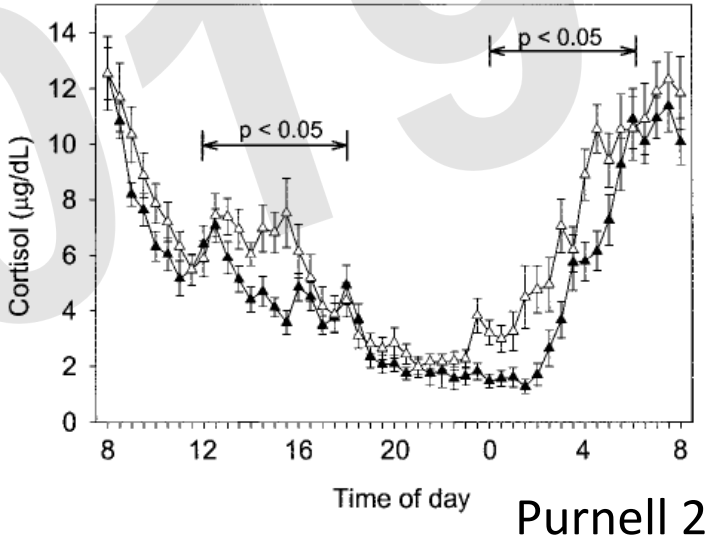
LOWS:

Na^+ (high K^+)

Glucose

Hemoglobin

Neutrophils



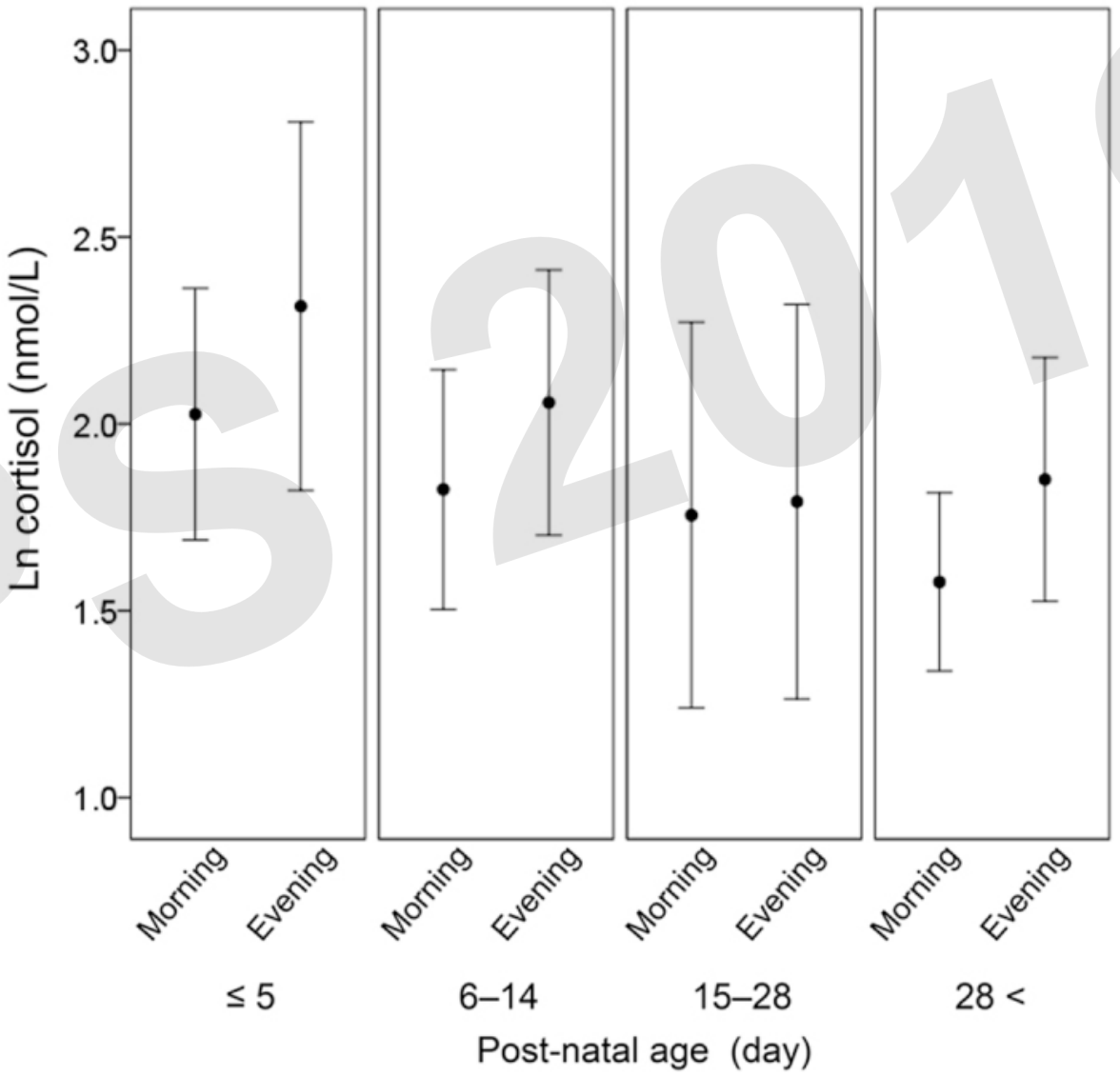
Purnell 2004

CORTISOL

(<9:00)

Neonates?

N=62



Kinoshita Sci Rep 2016

Management

Fluid resuscitation (NaCl 0,9% + glucose)

Glucocorticoids I.M. or I.V. : hydrocortisone
(or prednisolone)

25 – 50 – 100 mg/m² (>12y) bolus, then 6 hourly

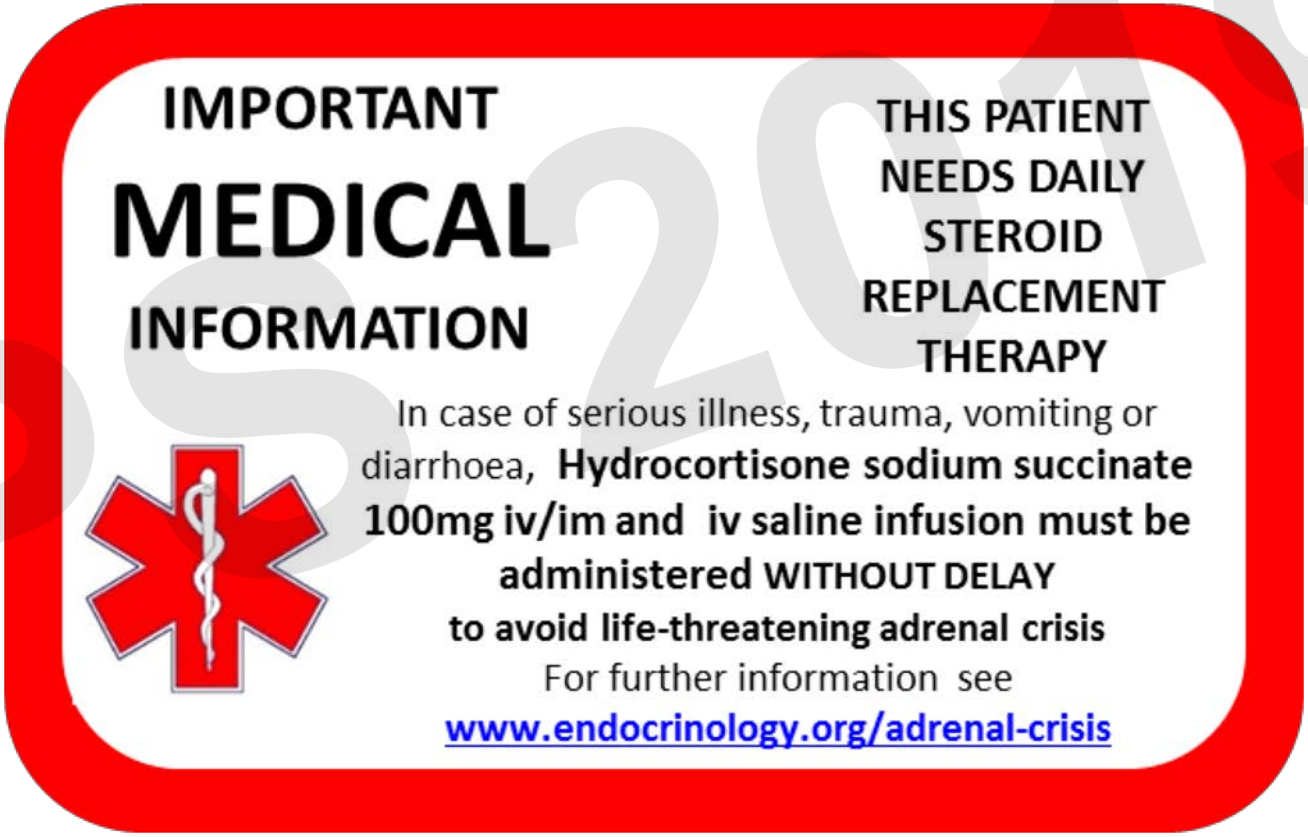
When >50 mg/d, fludrocortisone not required

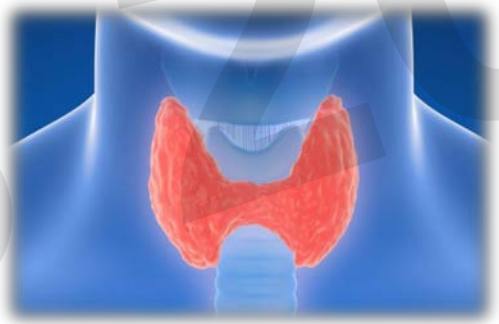
Prevention

Patient education

(80% of patients unable to adjust HC dose !)

Double or triple HC dose during illness, give stress dose before/during surgery


IMPORTANT
MEDICAL
INFORMATION
THIS PATIENT NEEDS DAILY STEROID REPLACEMENT THERAPY
In case of serious illness, trauma, vomiting or diarrhoea, Hydrocortisone sodium succinate 100mg iv/im and iv saline infusion must be administered WITHOUT DELAY to avoid life-threatening adrenal crisis
For further information see
www.endocrinology.org/adrenal-crisis



Neonatal Hyperthyroidism

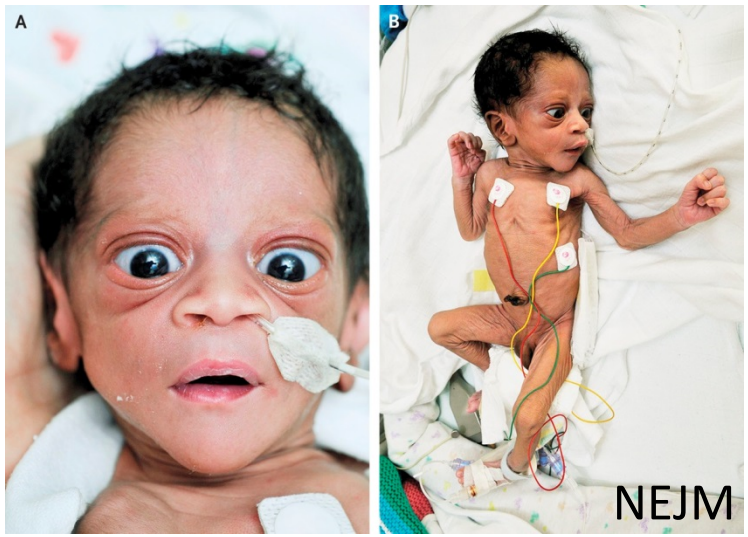
Maternal TSHR-SAb

Activating mutations of the TSH receptor
McCune Albright syndrome

1 in 1000 pregnancies is associated with Graves

Thyrotoxicosis in 1-5% of babies of mothers with Graves

1 in 25,000 neonates



Neonatal Thyrotoxicosis

Symptoms

Tremor, hypervigilance, irritability, poor sleep

Poor feeding, frantic suck, diarrhea, weight loss

Signs

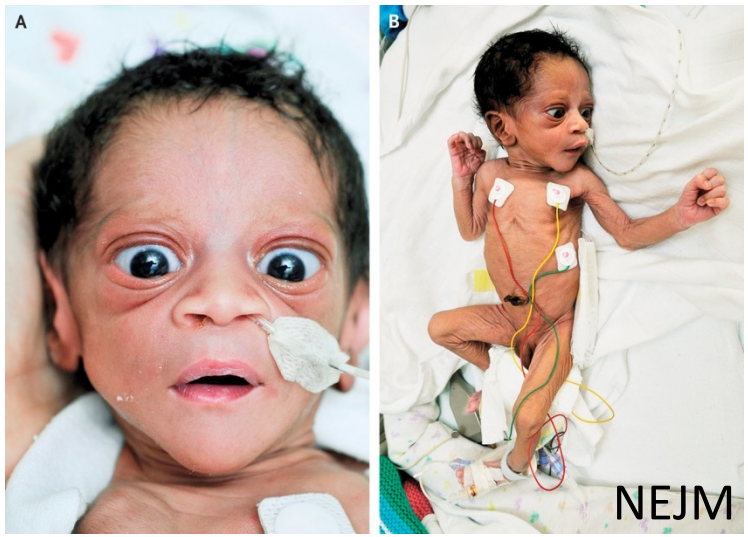
SGA, microcephaly

Thyrotoxic stare (adrenergic)

Warm and moist skin

Small goiter

Rapid heart rate, arrhythmias, heart failure



Neonatal Thyrotoxicosis

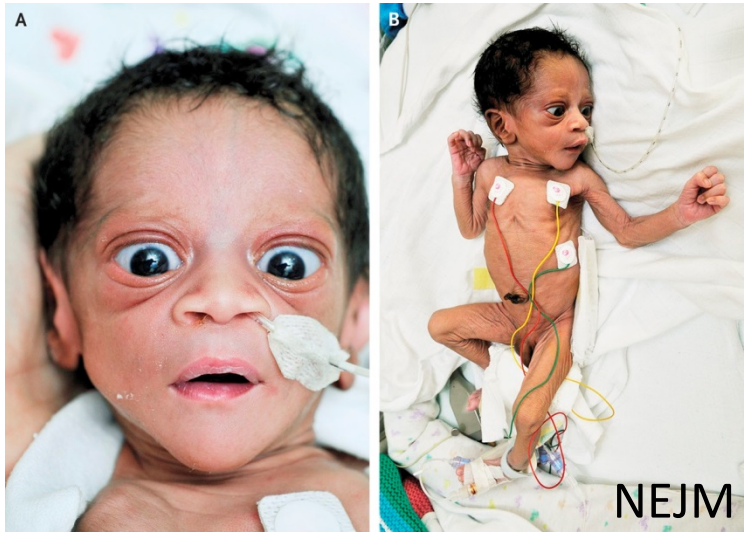
Fetal monitoring during pregnancy to detect fetal thyrotoxicosis:

- Goiter
- FHR (treat if sustained at > 160/min)
- Risk for heart failure, hydrops and IU demise

IU trt: PTU to mother to keep FHR at ~ 140

Maternal TSI titers: risk for neonatal thyrotoxicosis ↑ when

- TSI > 300% (very likely if > 500%)



May manifest after 4-5 days of birth

- PTU effects wear off
- ↑ conversion of T4 to T3

Up to 10-20 days after birth

Resolves in 3-12 w (Treatment necessary usually until it resolves)

Neonatal Thyrotoxicosis

Methimazole: Inhibits organification

- $T_{1/2}$ of 5-6 h
- 0,25-1,0 mg/kg per day
- Titrate when TSH levels increase >50th percentile

Iodine (used for 1-2 w): Inhibit organification and release of TH

Lugol's iodine (126 mg iodine/ml or 8 mg iodine/drop): 1-2 drops po qd to tid (preferred in neonates)

Beta Blockers (Propranolol; 2 mg/kg per day q4)

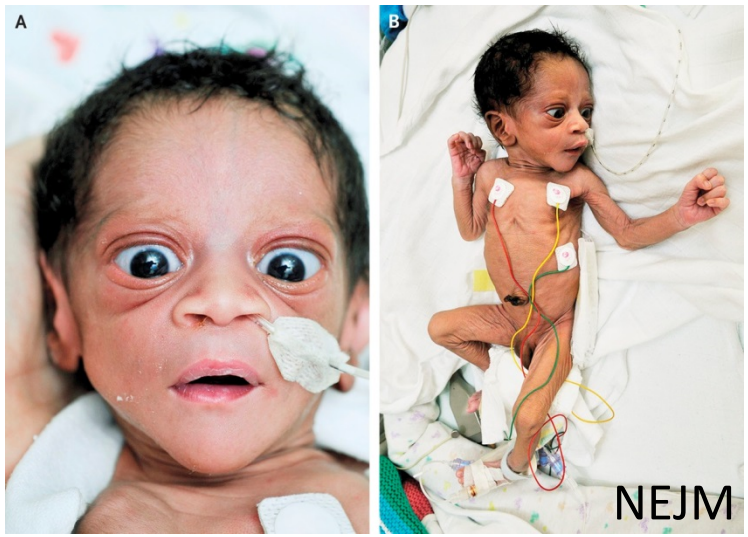
Glucocorticoids: ↓ T4 secretion and conversion of T4 →T3

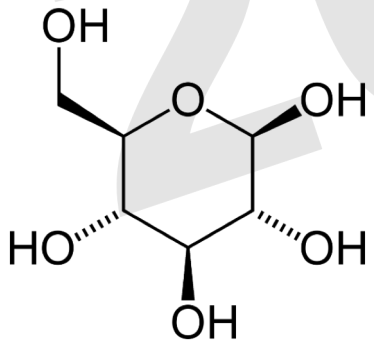
Neonatal Thyrotoxicosis

TSH may remain low for several weeks after resolution: requires T4 replacement

F/U:

- Poor growth
- Craniosynostosis
- Hyperactivity
- Developmental and behavioral problems described





♂ 5 do, **neonatal dizziness**

d1: hypoglycemia 13 mg/dL

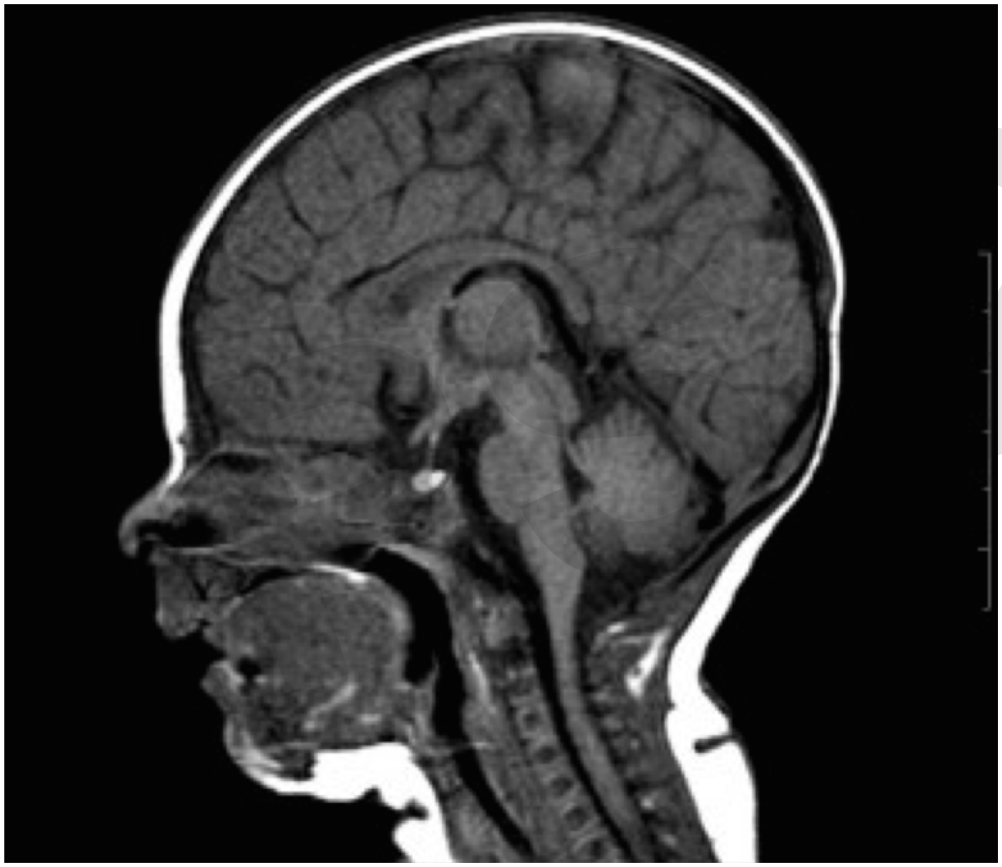
d2: hypoglycemia 20 mg/dL

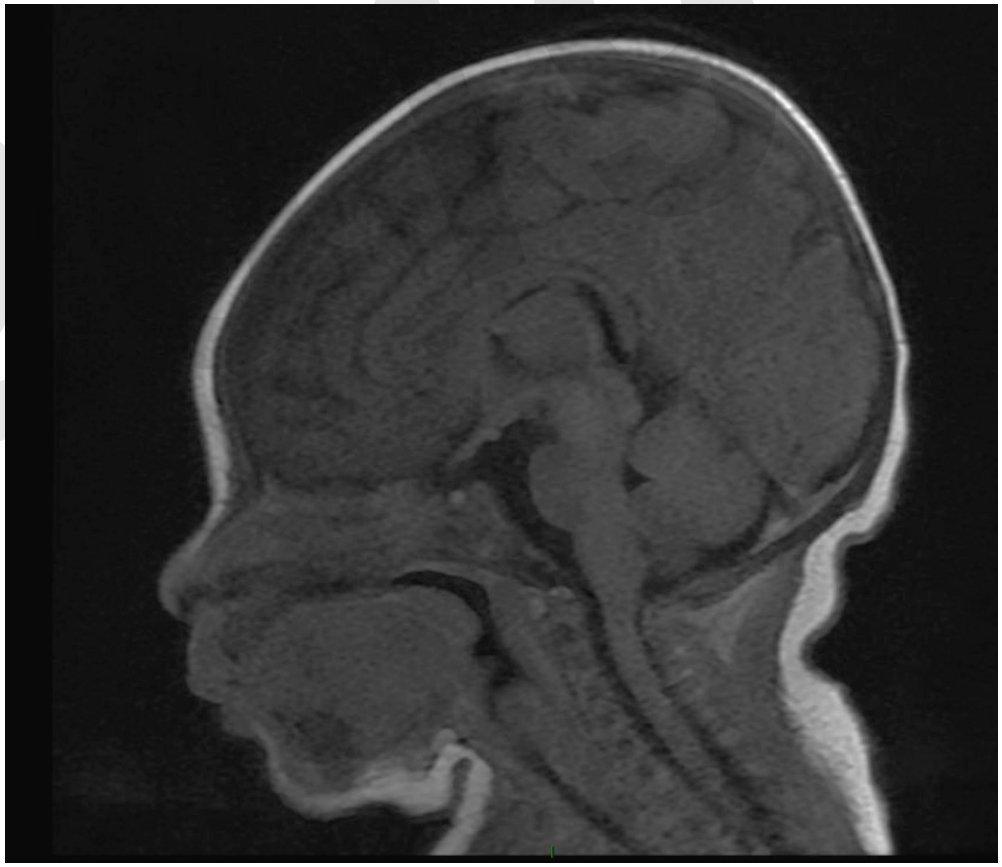
Glycemia	49 mg/dL
Venous pH	7,34
Lactate	0,7 mmol/L
HC03-	24 mmol/L
Insulin	0,26 µUI/mL
Cortisol	52 nM
GH	1,8 ng/mL
Na+	139 mmol/L
K+	4,84 mmol/L
GPT	25 U/L

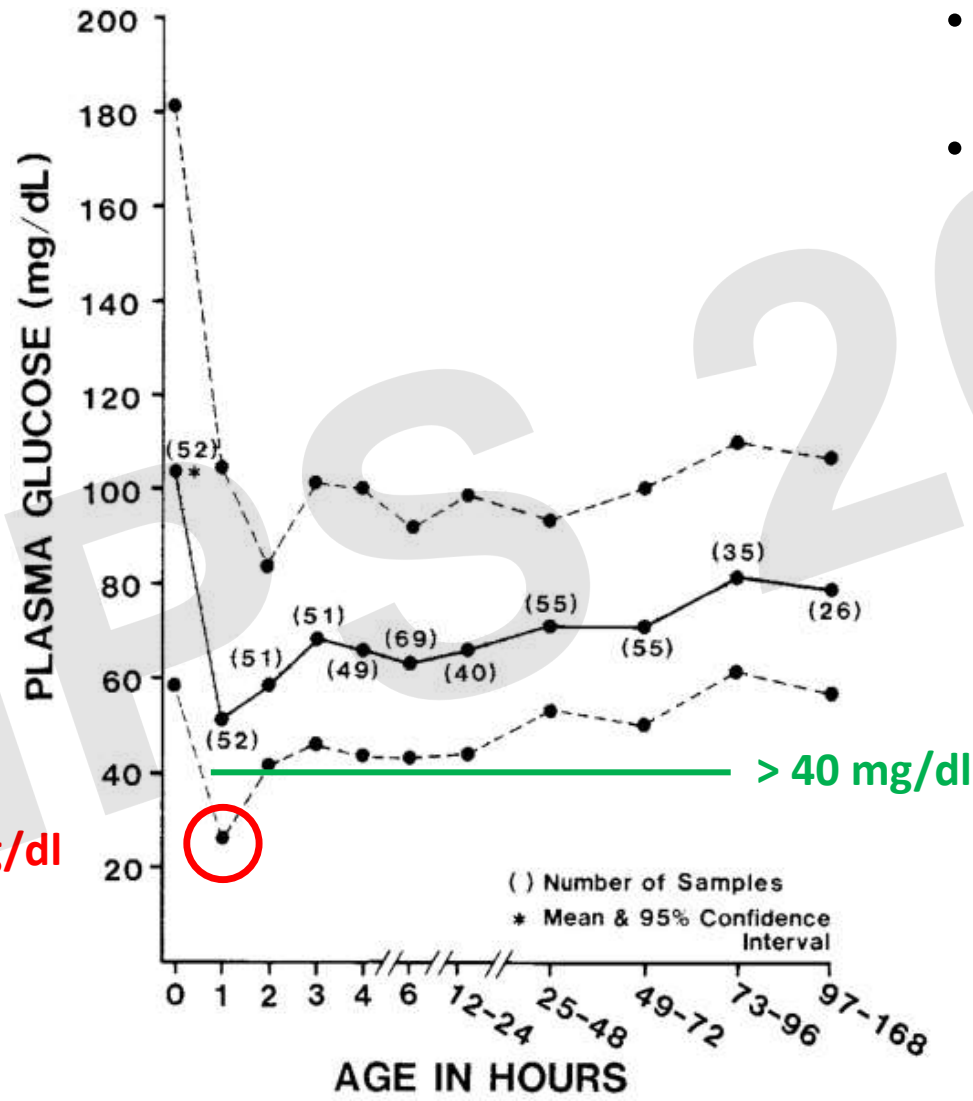
Complementary endocrine tests:

TSH	1,69	mU/L (1,36 - 8,76)	at d6 ↘
T4L	11	pmol/L (10,8 - 20,3)	at d6 ↘
ACTH	4,7	pg/mL (5-49)	↘
IGF1	24,5	ng/mL (24-117)	

PANHYPOPITUITARISM







(J. Pediatrics, 1996)

- Physiological nadir of blood glucose
- BG value needs to be confronted to timing and sort of dosage (plasma vs strips)

When to perform BG screening?

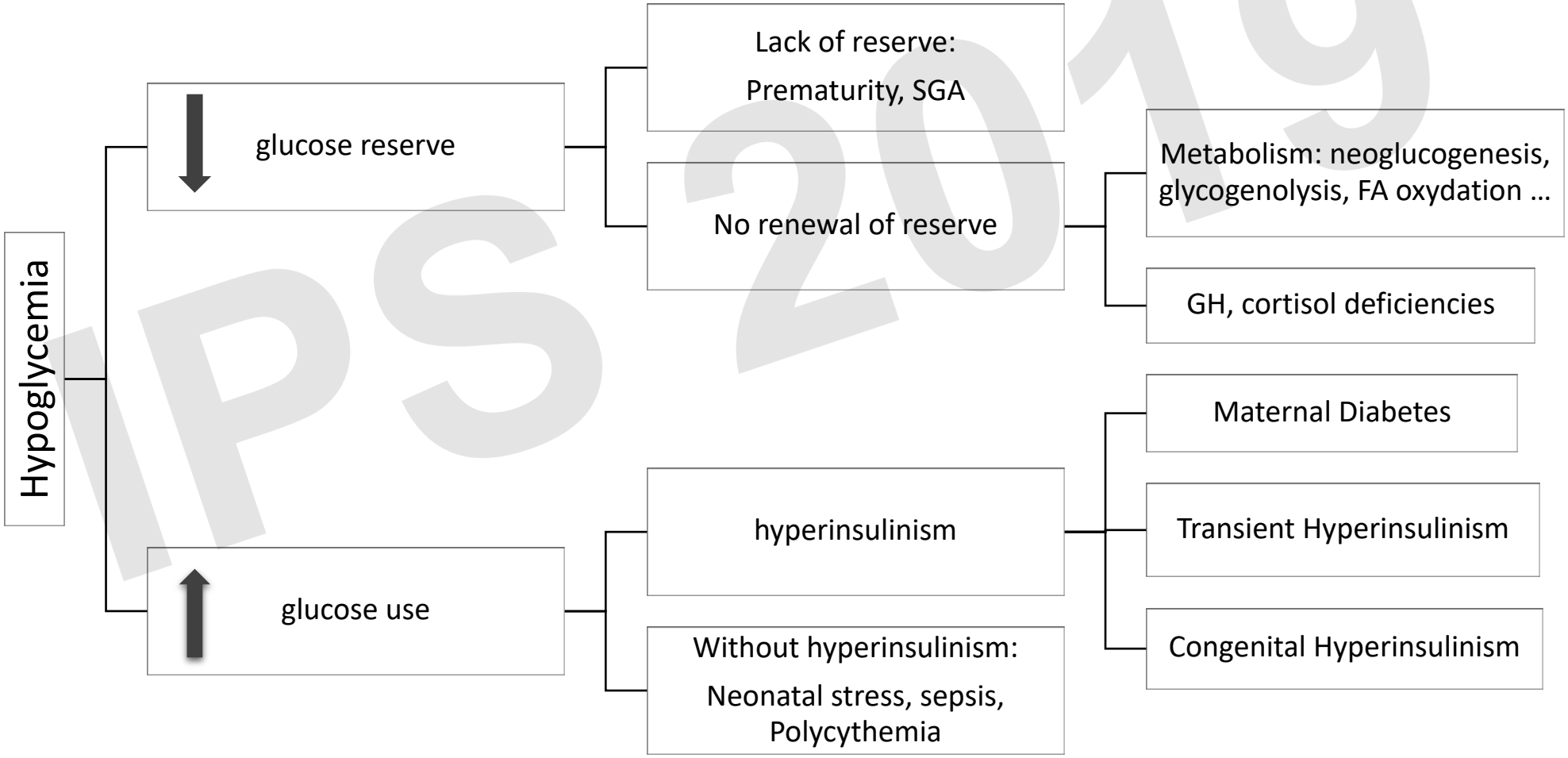
- Symptomatic newborns

- Screening < 1h of life in at-risk newborns:
  - **« Late » prematurity**
  - **SGA (<P10)**
  - **Diabetic mother**
  - **LGA (>P90)**
  - Postmature
  - Beta-blockers
  - Perinatal stress (asphyxia, hypothermia, sepsis)

NO screening in newborns at term, eutrophic, and asymptomatic!

D.H. Adamkin, march 2011, *AAP*

Etiologies



Work up

WHEN?

Every hypoglycemia persisting >48-72h

OR

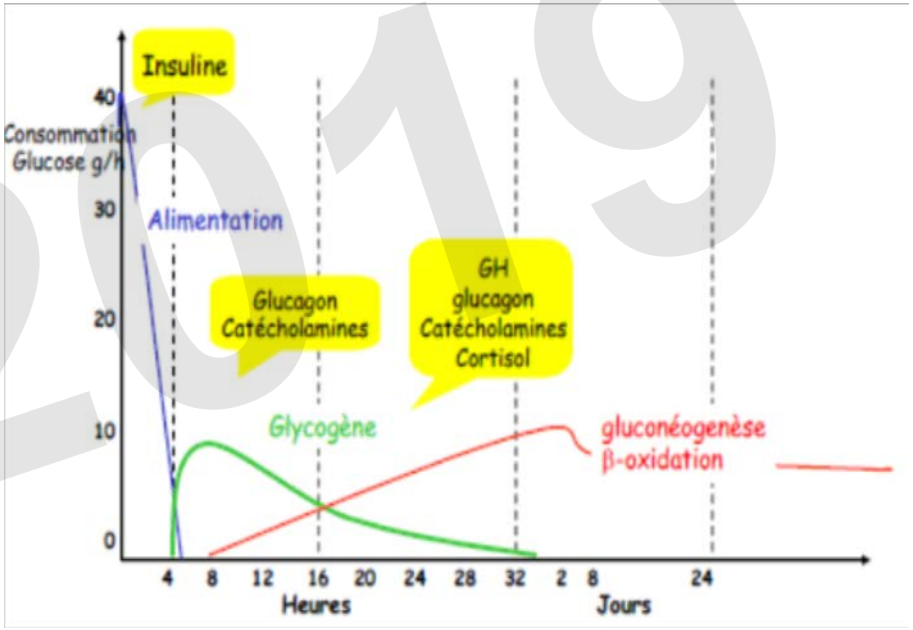
Precocious Hypoglycemia: brutal, profond and highly symptomatic

Work up

BLOOD:
- Glycemia
- Venous pH, lactate, HCO3-
- Insulin, GH, cortisol
- Electrolytes, GOT, GPT

URINES:
- Ketone bodies

- Acylcarnitins
- Ammonium (on ice)
- Ketones (BOHB), FFA -> Tubes at -70° C
- C-peptide
- Urinary organic acids



1. **Timing** of hypoglycemia
2. **Hepatomegaly**? Micropenis, cleft palate, hyperpigmentation?

Endocrine treatment

T4: urgent… but need to ensure GC supplementation!

50 µg/kg/d

Give before d15

Titrate after 3w

GC: hydrocortisone: newborns need 15-20 mg/m²/d

(3q or 4q)

Growth hormone: early treatment is recommanded

(neurological development)

Consequences?

Symptomatic hypoglycemia:

Burns; *Pediatrics* 2008: Prospective study

35 newborns at term with symptomatic hypoglycemia

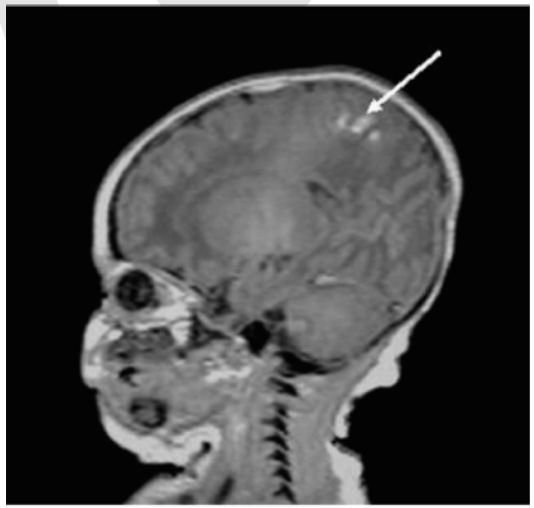
22 transient

13 prolongated/recurrent

-> **94%** of anomalies of **brain white matter** at MRI

-> **65%** of developmental retardation at 18 months of age

(Burns; *Pediatrics* 2008)



Asymptomatic hypoglycemia**:** controversed

- At risk groups may have bad neurological outcome

- Does the efficacy of hypoglycemia treatment/prevention influence the outcome?

(Mckinlay 2018, Kaiser 2015)

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Hasnae Boughaleb
Juliette Vanderroost

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Treatment

SYMPTOMATIC

- IV Glucose

  Bolus Dextrose 10% 2ml/kg in 5 min (=200mg/kg)

  + perfusion of glucose 5-8 mg/kg/min

- Glucagon

  If persistence of hypoglycemia while glucose perfusion
  8mg/kg/min

  Dose: 20-30 µg/kg in IV of SC

ASYMPTOMATIC

- Oral

  Breastfeeding or milk <1h

  Dextrose gel 40%

- IV Glucose

  Continuous perfusion of glucose

  5-8 mg/kg/min

  (NO BOLUS)