

BELGIAN PRIMARY ADRENAL INSUFFICIENCY REGISTRY (PAI-BEL)

Center number		Patient identification (2 last letters of first and last name)	
Patient number			
Name of the physician		Date of last clinical visit	

II] CONGENITAL ADRENAL HYPERPLASIA (CAH) FORM

1) Patient Information

Birth data (mm/yyyy)

Gender (M/F)

Ethnicity (Caucasian/North-African/African/Asian/Other/Unk)

First language

Postal Code

Marital Status

Single		Married		Cohabiting		Widower		Divorced		Unknown	
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Professional activity

Full time		Part time		Disability		Pensioner		Job seeker		Student		Unknown	
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Year of diagnosis (yyyy)

Status at enrolment

Alive		Died	
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If died at enrolment

Date		Cause	
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Menopausal status

Yes		No		If yes, age		Unknown	
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Tobacco use

Yes		No		Unknown	
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2) Diagnostic Circumstances of CAH (check, several possibilities)

Peri-natal period		Ambiguous genitalia		Dehydration	
Hyponatremia		Hyperkalemia		Hypovolemia	
Growth failure		Weight loss		Pseudo-precocious puberty	
Hypoglycemia		Adrenal crisis		Other (specify):	

3) Molecular testing (if known)

Gene	Result
STAR	
HSD3B	
CYP21	
CYP11B	
Other	

In case of classic 21-hydroxylase deficiency:

Salt-wasting form		Simple virilizing form	
Mutation (if known):			

4) Test Confirming Diagnosis (check, several possibilities)

	Value	Units	Normal range
Serum cortisol 8 AM			
Serum ACTH 8 AM			
Serum aldosterone			
Serum renin (direct/activity)			
17OH-Progesterone			
11-deoxycortisol			
17OH-Pregnenolone			
Δ4-Androstenedione			
DHEAs			
SYNACTHEN® (250 µg) Basal cortisol Stimulated cortisol (60') Basal 17OHP Stimulated 17OHP (60')			
Other test (precise which)			

5) Concomitant Diseases

Type of disease	Yes	No	Unknown
Diabetes			
Hypertension			
Osteoporosis			
Hyperlipidemia			
If other, precise:			

6) Medical Familial History

Yes	No	Unknown
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Family relationship, specify	Type of disease

7) Fertility Information

For Men

Paternity:

Yes	No	Unknown	Not relevant
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If known, number of children:

Details (if known):

	Number of children
Spontaneously	
Men assisted reproduction	
Adoption	

Echography for Testicular Adrenal Rest Monitoring:

Yes	No	Unknown
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If yes,

Date of last result	Result

Cryopreservation:

Yes	No	Unknown	Not relevant
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For Women

Genital surgery:

Yes	No	Unknown	Not relevant
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Gravida:

Yes	No	Unknown	Not relevant
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If known, number of children:

Details (if known):

	Number of children
Spontaneously	
Women assisted reproduction	
Adoption	

Other details, if known:

Precise for each gravida					
Year of gravida					
Delay between desire and conception (months)					
Pregnancy outcome Singleton, Twins, Triplets Live Birth Stillborn Elective Termination Spontaneous abortion Gestational age at spontaneous abortion	Specify or Check:				
	Yes / No	Yes / No	Yes / No	Yes / No	Yes / No
	Yes / No	Yes / No	Yes / No	Yes / No	Yes / No
	Yes / No	Yes / No	Yes / No	Yes / No	Yes / No
	Yes / No	Yes / No	Yes / No	Yes / No	Yes / No
Delivery Date					
Caesarian section	Yes / No	Yes / No	Yes / No	Yes / No	Yes / No
Gestational Age at delivery >42 weeks (post mature) 38-42 weeks (term) <38 weeks (preterm)	Check:				
Were there any complications during the pregnancy? Specify:	Yes / No	Yes / No	Yes / No	Yes / No	Yes / No
Were there any perinatal complications? Specify:	Yes / No	Yes / No	Yes / No	Yes / No	Yes / No
Were there any Congenital Abnormalities Specify:					
Change in glucocorticoid replacement during pregnancy Precise:	Yes / No	Yes / No	Yes / No	Yes / No	Yes / No
Pre-Pregnancy Weight (kg)					
End-pregnancy Weight (kg)					
Baby Weight at birth (kg)					

8) Adrenal Crisis Events

Did the patient experience an adrenal crisis event ?

Unknown	Never	Yes	If yes, how many during 2 last years	
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If yes, precise for each crisis:

Date				
Circumstances				
Hospitalisation (yes/no) duration				

9) Rate of medical visits/hospitalisations

During the year before last visit, was the patient hospitalized ?

Unknown	No	Yes	If yes,	how many times	
				reason	

10) Last Patient Visit

Date

Physical Examination

Height (cm) Current Weight (kg) Blood Pressure (mmHg) Heart Rate (beats/min)

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Check if positive:

Facial plethora		Dorsocervical fat pad		Supraclavicular fullness	
Centripetal obesity		Abdominal striae		Weakness, proximal myopathy	
Bruising		Ankle edema		Hirsutism / Acne / Oily skin	
Dysmenorrhea		Hypertension		Other:	

Laboratory Assessments

	Value	Units	Date
Fasting plasma glucose			
HbA1c			
Total Cholesterol			
LDL-cholesterol			
HDL-cholesterol			
Triglycerides			

Dual Energy X-ray Absorptiometry (DEXA)

Yes	No	Unknown	Not relevant
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Date of last result	Result	
	T-score lumbar spine	Z-score lumbar spine
	T-score femoral neck	Z-score femoral neck

11) Current Treatment**Glucocorticoid Replacement Therapy**

Yes	No
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Date of beginning replacement therapy Type (check):
If other:

Hydrocortisone	Prednisolone	Cortisone acetate	Dexamethasone	Other
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Timing	Morning	Midday	Afternoon	Evening	Bedtime	Overnight
Dose (mg)						
Total daily dose (mg)						

Reason for this scheme (check, several possibilities):

Medical advice	Night worker	Patient choice	Lifestyle	Poor compliance	other
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Mineralocorticoid Replacement Therapy

Yes	No
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Date of beginning replacement therapy

Timing	Morning	Midday	Afternoon	Evening	Bedtime	Overnight
Dose (μg)						
Total daily dose (μg)						

Use Parameters for Gluco- and/or Mineralo-Corticoid Adaptation

Yes	No
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If yes, specify	
Clinical	
Biological	
Other	

Other Concomitant Medications

Name	Dosage (U)	Frequency	Ongoing

12) Educational Information to Prevent Adrenal Crisis (check, several possibilities)

General information about disease		Information related to recognition of adrenal insufficiency signs		Instructions for adaptation of replacement therapy	
Prescription of Soluortef®		Adrenal insufficiency card / bracelet / necklet		Patient referred to disease support group	

If other, specify:

Educated person (check, several possibilities)

Patient		Family		Family doctor		Other	
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If other, specify:

Educational information made by (check, several possibilities)

Endocrinologist		Nurse		Family doctor		Other	
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If other, specify: