CHARACTERISTICS OF CENTRAL PRECOCIOUS PUBERTY CAUSED BY HYPOTHALAMIC HARMATOMA

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INTRODUCTION

- Hypothalamic hamartomas (HH)
- ✓ rare congenital malformations (1-2 /100 000 incidence)
- ✓ Benign
- ✓ Consequences:
  - precocious puberty,
  - recognition deficiencies
  - gelastic seizures
  - behavior disorder

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INTRODUCTION

- Precocious puberty due to harmatoma starts at very young age
- If no treatment it can affect:

  - Height growth
  - Serious health problems
  - Psychological crisis

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Contents

Precocious puberty caused by harmatoma

- *Clinical features*
- Investigations

Subjects and methods

1. Subjects

- 16 patients with Hypothalamic harmatoma
- Endocrinology department, the national hospital of pediatrics
- Time: 2000 – 2016
- Diagnosis criteria by Carel 2008

Carel JC, Leger J (2008). Precocious Puberty.*N Eng J Med*; **358 (22)**: 2366-2377

Subjects and methods

2. Methods

- A retrospective study
- Secondary sexual characteristics by Marshall and Tanner
- Bon age: Xray of the left wrist
- FSH, LH, estradiol in girls, testosterone in boys.
- GnRH stimulation test
- Skull MRI: harmatoma.
- Abdominal ultrasound to exclude other causes

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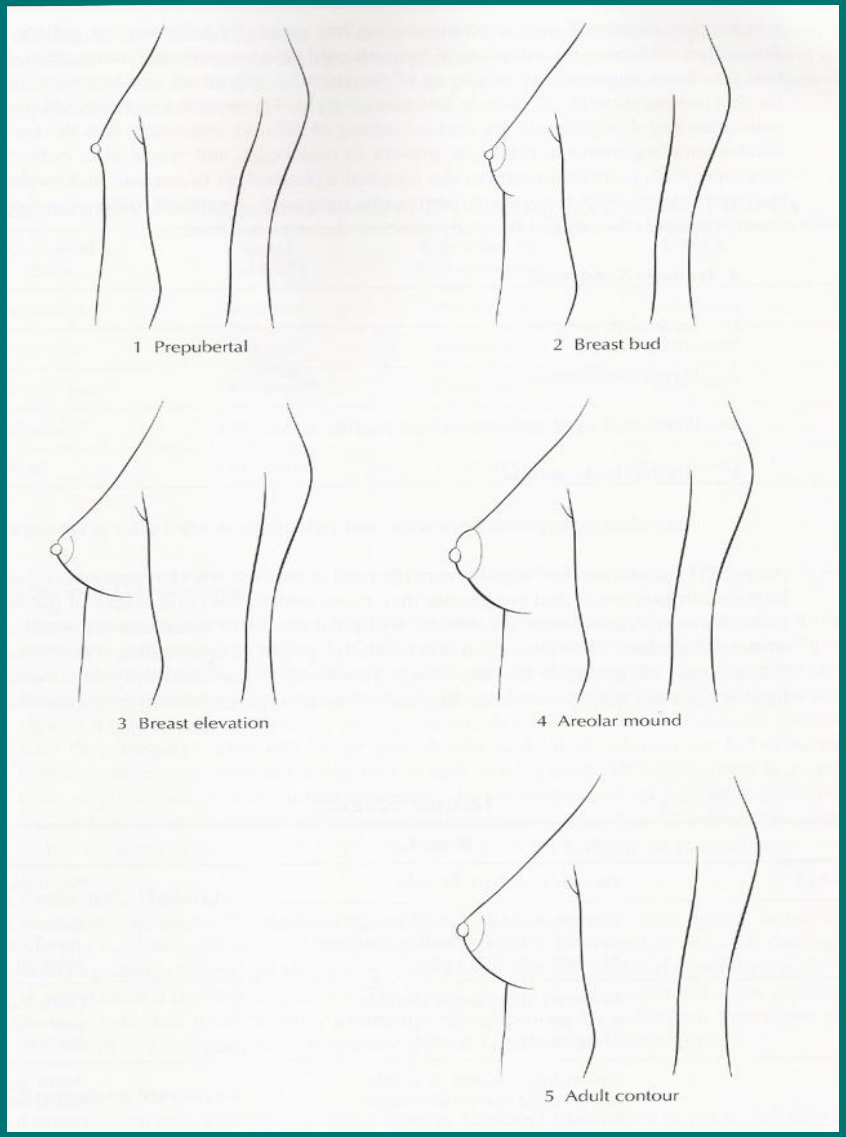
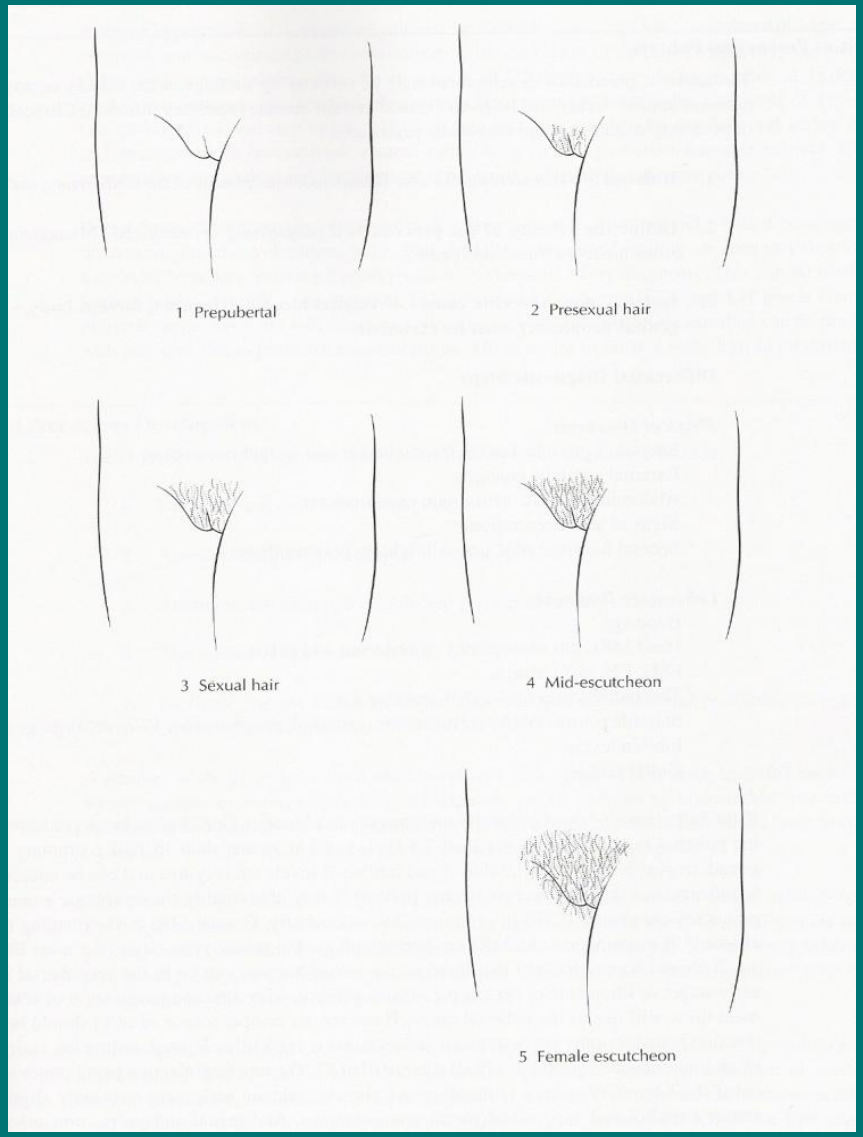
TANNER Stages





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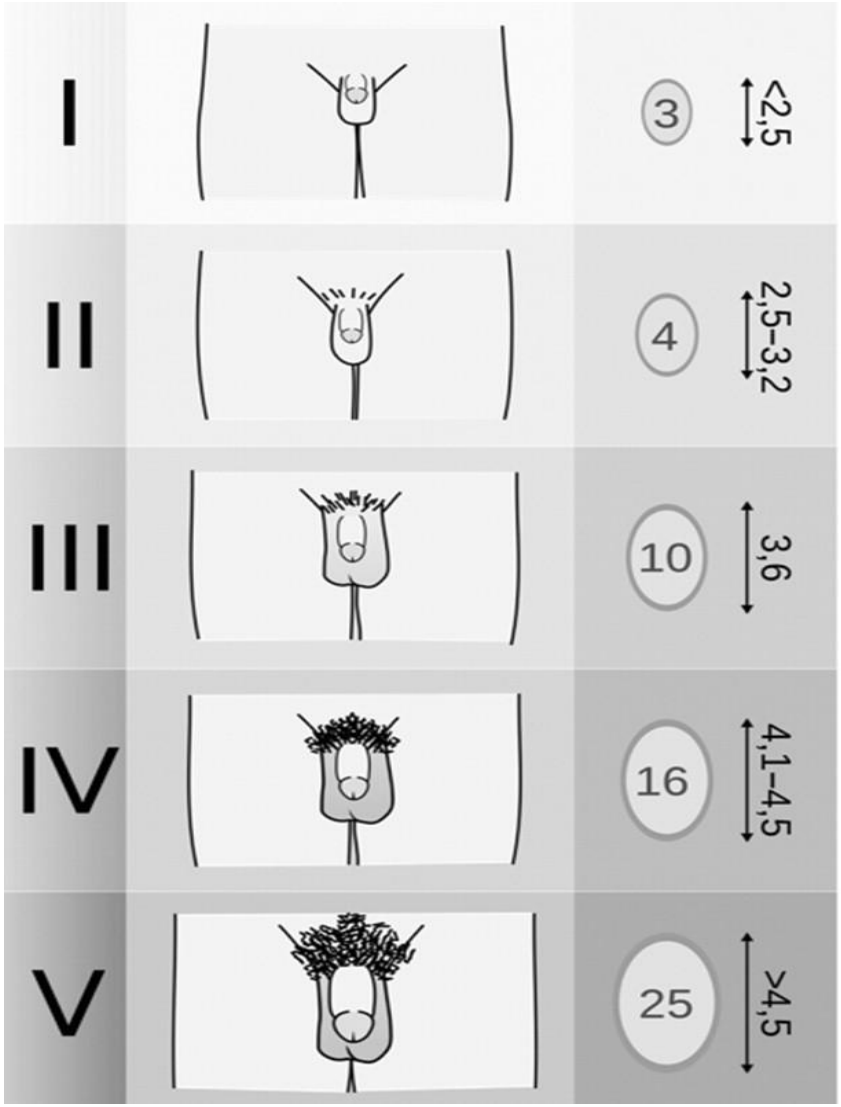
TANNER Stages



Stage I: prepubertal; testicular size less than 4 cc in volume and 2.5 cm in longest dimension

Stage II: enlargement of scrotum and testes; scrotal skin reddens and changes in texture; growth of testes to 4 cc or greater in volume

Stage III: enlargement of penis (length at first); further growth of testes

Stage IV: increased size of penis with growth in breadth and development of glans; testes and scrotum larger, scrotal skin darker

Stage V: adult genitalia

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Results

❖ Ages of diagnosis

- ✓ Boys: 15 - 96 months (average 55,8 ± 11,2 months)
- ✓ Girls: 19 - 96 months (average 46,1±9,3 months)

❖ Chief complains:

- ✓ Boys: penis growths (100%)
- ✓ Girls: breast development 62,5%, vaginal bleeding 37,5%

❖ Signs and symptoms started before exams

- ✓ Boys: 7,3±2,1 months
- ✓ Girls: 11,3±3,7 months

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Results

Clinical features in girls

Characteristics	Stages by Tanner	n
Breast	B1	0 (0%)
	B2	3 (37,5%)
	B3	4 (50%)
	B4	12,5 (0%)
	B5	0 (0%)
Public hair	P1	6 (75%)
	P2	2 (25%)
	P3	0
	P4	0
	P5	0
Menarche	3 (37,5%)	
Acne	0 (0%)	

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Results

Clinical features in boys

Characteristics	Results	n
Penile length	7,1±1,7 (cm)	8
Testicular thickness	10,1±4,3 (cm)	8
Acne		2 (25%)
Public hair	P1	5 (62,5%)
	P2	2(25%)
	P3	1(12,5%)
	P4	0
	P5	0

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Clinical features

Picture 1. A 2 year boy, 4ml testicular and 8 cm penile length

Picture 2. A 2,5 year girl with breast of B3 clitoris growth

4 years, central precocious puberty due to harmatoma

Investigations

Girls

	Day of diagnosis	3 months after treatment	N	P
LH (UI/L)	5,4±2,2	0,5±0,2	8	
FSH (UI/L)	6,4±2,2	1,4±1,0	8	<0,05
Estradiol (pmol/L)	168,5±63,4	24,7±9,5	8	

Boys

	Day of diagnosis	3 months after treatment	N	P
LH (UI/L)	2,4±0,6	1,2±0,6	8	>0,05
FSH (UI/L)	8,8±3,7	0,69±0,3	8	
Testosteron (nmol/L)	17,4±5,1	0,45±0,3	8	<0,05

Hormone increased equivalent to puberty level on the day of diagnosis

3 months after treatment by GnRH analogs, the hormone level decreased ($p < 0,05$)

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Imaging diagnosis

❖ ***Boys:***

- Normal testicular untrasound.
- Average bone age: 93,0± 42,9 months, higher than real age 34,5±15,7 months.
- Skull MRI found a hypothalamic harmatoma Of 12,2±6,6mm x 14,8±6,5 mm.

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Imaging diagnosis

- ***Girls:***

- Uterus ultrasound:size of 11,3±3,7mm x 42,2±5,8 mm.
- Average bone age: 67,5 ±28,6 months, higher than chronological age 23,8±11,3 months.
- MRI of skull found a harmatoma in the thalamus, Average size 10,8±0,8mm x 13,8±1,2 mm.


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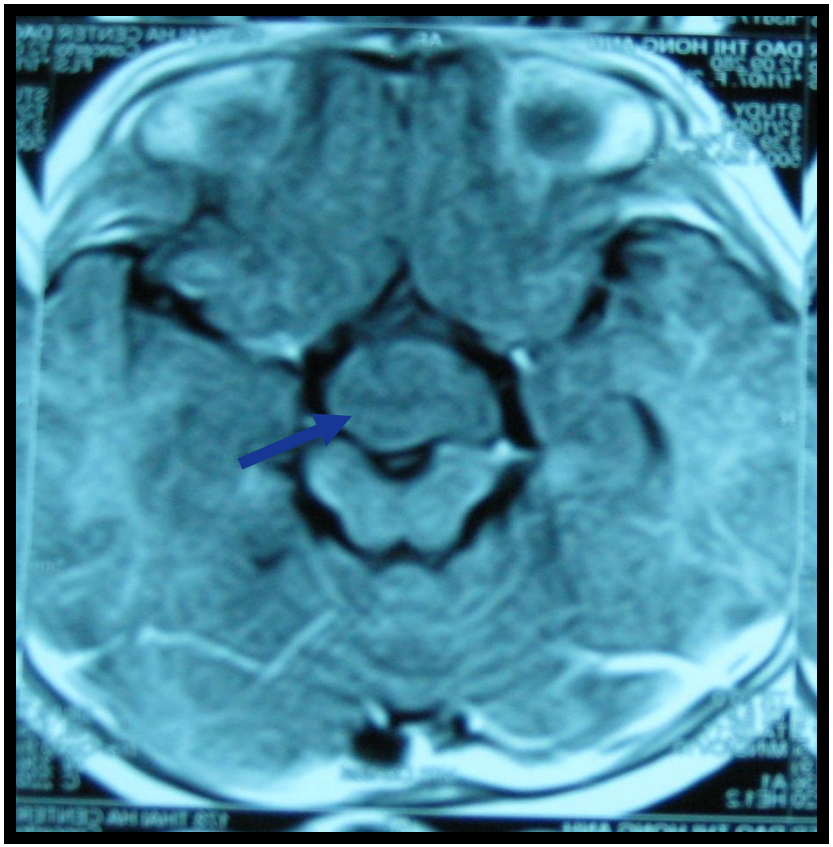

- 5 year girl
- Investigations
- FSH: 6,98 UI/l
- LH: 6,29 UI/l
- Estradiol: 0,32 pmol/l
- Bone age: 7 tuổi

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Conclusion

- ✓ Harmatoma: a cause of precocious puberty
- ✓ Specific clinical features
- ✓ Increase bone age dramatically
- ✓ Sexual hormone and gonadotropin increase significantly
- ✓ Skull MRI is essential to diagnose

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