



Impaired growth hormone secretion associated with large hypothalamic hamartoma

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Disclosure Statement: there is no conflict of interest

Background

Hypothalamic hamartomas (HH) are rare non-progressive brain tumor malformations that occur early during brain development. Most of the cases are sporadic and nonsyndromic. Variation in size and location of the hamartoma within the hypothalamic region is not always associated with the severity of the clinical presentation.

A wide range of symptoms could occur - from nonsymptomatic to severely affected cases that include seizures (mostly gelastic), behavioral difficulties and endocrinological disturbances.

Among all, precocious puberty is most common endocrinological dysfunction; others present less frequently.

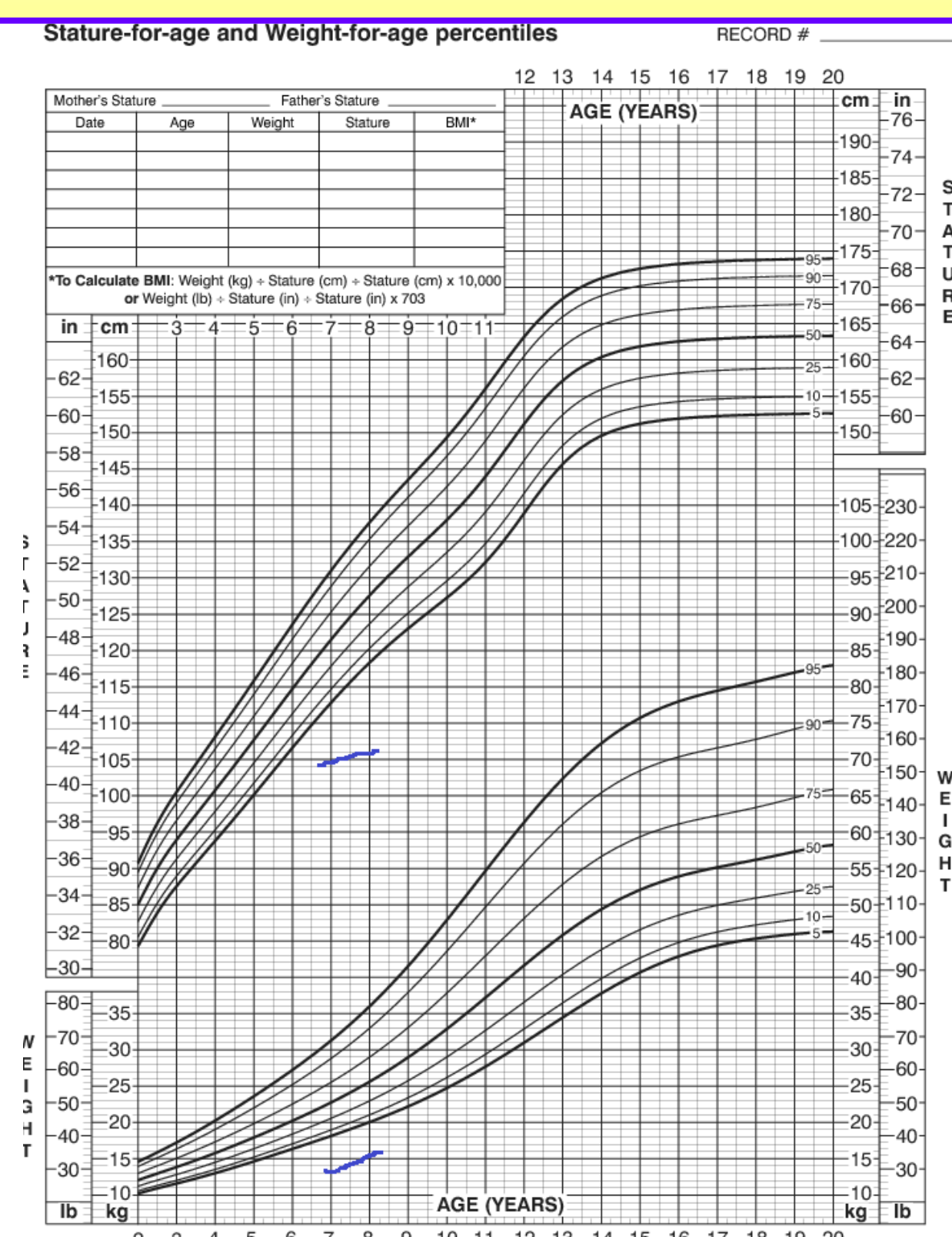
Materials and methods

A girl approached our clinic due to the short stature. The child was born on term BW3350gr/BL50cm. During early infancy growth was within the normal curve.

Growth velocity started to decline after the age of 4 years.

At the age of 7 years the child was 4SDS below the mean for height and 3SDS for the weight.

Pubertal development started at 8,5 years of age.



Growth curve showing declining of the growth velocity

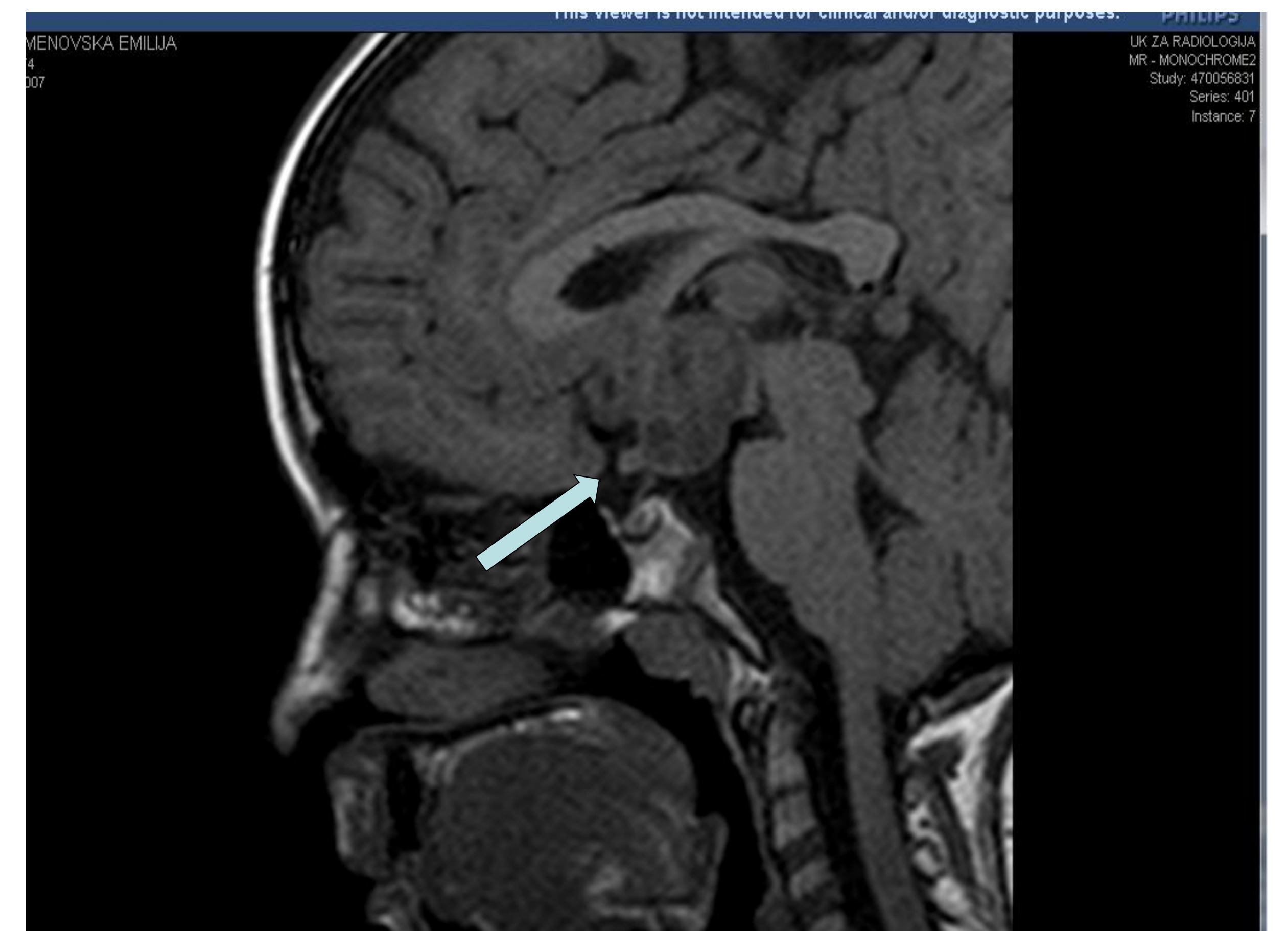
Results

Endocrinologic data

- Wrist X ray - delayed maturation for 2 years
- GH deficiency was confirmed with stimulation tests (peak value of 4,3 ng/ml)
- low IGF1
- other pituitary hormones were normal for the age

Neurologic data

- behavioral problems -shyness, timidity and inability to accustom to school activities
- frequent headaches
- episodes of crying
- normal neurological examination and ortooptic testing
- normal intelligence



MRI of the pituitary showed large hypothalamic hamartoma 30x28 mm without tendency for growth during the two following years. The structure and size of the pituitary gland was normal.

Conclusions

Clinical presentation of HH is mostly associated with a particular position of the hamartoma. Since the size of HH in our case is 30mm, both possible mechanisms could be involved - impaired GHRH secretion from the lower parts of the hypothalamus (arcuate nucleus) and inadequate control of somatostatin levels produced from upper parts of the hypothalamus (rostral periventricular nucleus). However, short stature should be considered as a rare feature of HH.