Increased required dose of growth hormone in children with inborn panhypopituitarism

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Conclusion:

In patients with inborn panhypopituitarism:

- **Higher doses of growth hormone are (initially) necessary to achieve appropriate growth velocity & normal IGF1-levels**
- **Therapy initiation with a dose of 35µg/kg body weight seems to be appropriate**
- **Close monitoring and adaption of GH dose is necessary**
- **IGF 1 levels and growth velocity should be in the upper normal range**

Introduction

Standard dose for growth hormone (GH) deficient children is 25 µg/kg/d given subcutaneously once daily. Inborn panhypopituitarism is a special subset of GH deficiency. Its management is difficult because several hormones need to be replaced. We present 3 patients with perinatal onset panhypopituitarism.

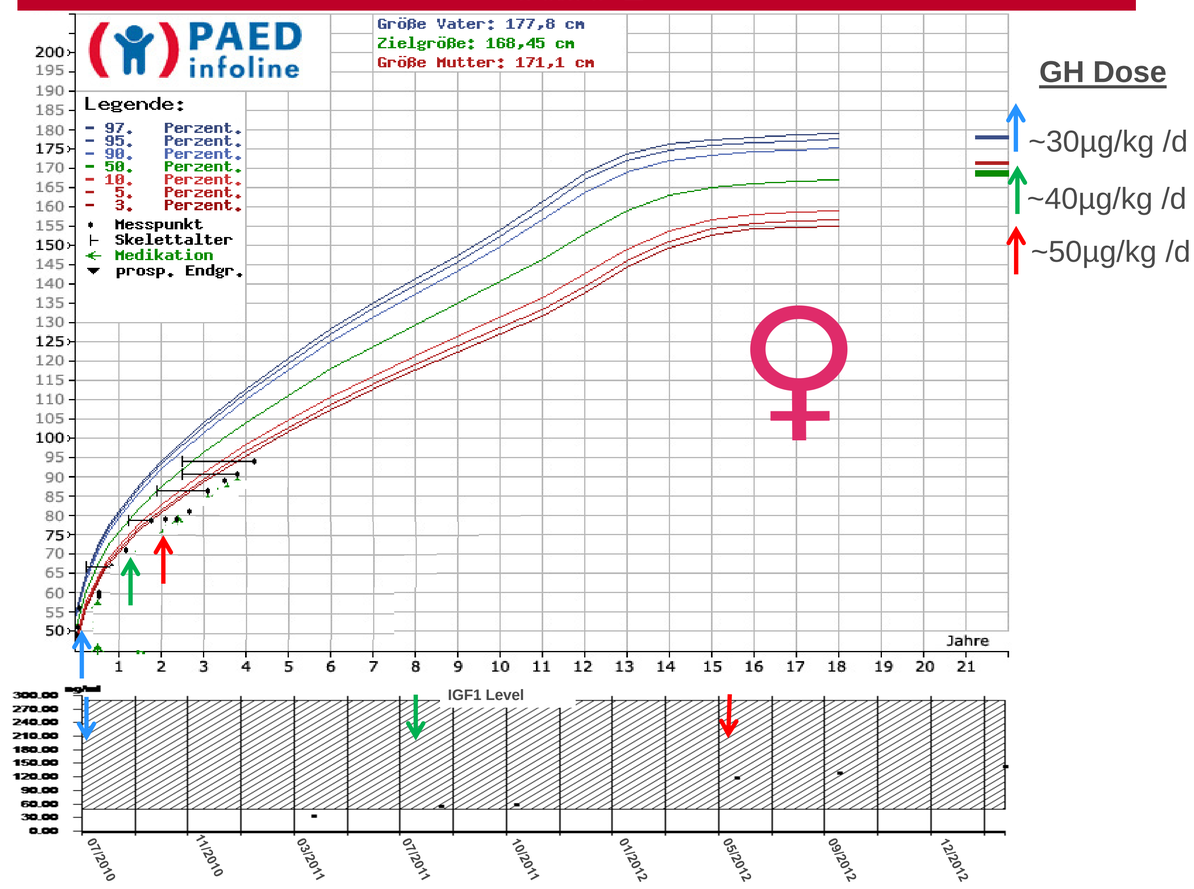
Case Reports

Case 1 is a 4-year-old girl who was born after fetal hydrops and emergency C-section. In the first few weeks of life hypothyroidism and hypocortisolism was diagnosed and treatment initiated. MRI showed aplastic hypophysis, hormonal testing revealed panhypopituitarism. Treatment with GH (25 µg/kg/d) was started, but no catch-up growth was reached and IGF1 remained low. GH-dose was increased in several steps to a dose of 52 µg/kg/d to achieve good growth velocity and IGF1-levels within the normal range.

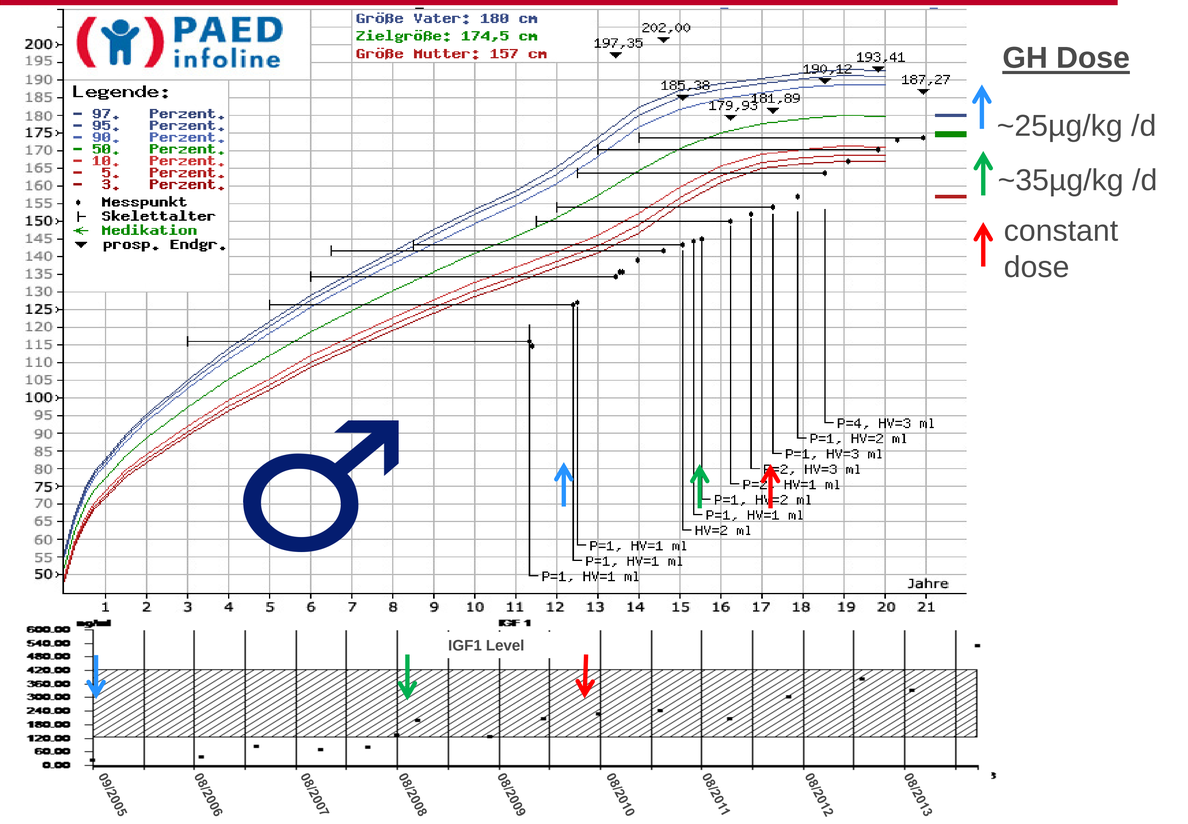
Case 2 is a 20-year-old man with hypoplastic and dystopic pituitary gland of unknown cause. Genetic testing was unsuccessful. Treatment for panhypopituitarism was initiated at the age of 11. He was treated with GH in a dose of 25 µg/kg/d, but catch up-growth and normal IGF1-levels were only achieved with a dose of 35 µg/kg/d. Despite his age the patient is still growing due to constitutional delay of growth and will reach his genetic length.

Case 3 is a 7-year-old boy who had hypoglycaemic seizure with reanimation due to circulatory arrest shortly after birth. Panhypopituitarism was diagnosed as hypoglycemia was ongoing during intensive care treatment. After initiation of treatment with GH and cortisol hypoglycemia ceased. MRI showed hypoplastic pituitary gland. GH therapy was increased up to a dose of 36 µg/kg/d to achieve good growth velocity and IGF1 in the upper normal range. Since the age of 4, dose/kg was slowly reduced to 26 µg/kg/d maintaining good growth velocity and good IGF1-levels. The patient’s prospective length is normal.

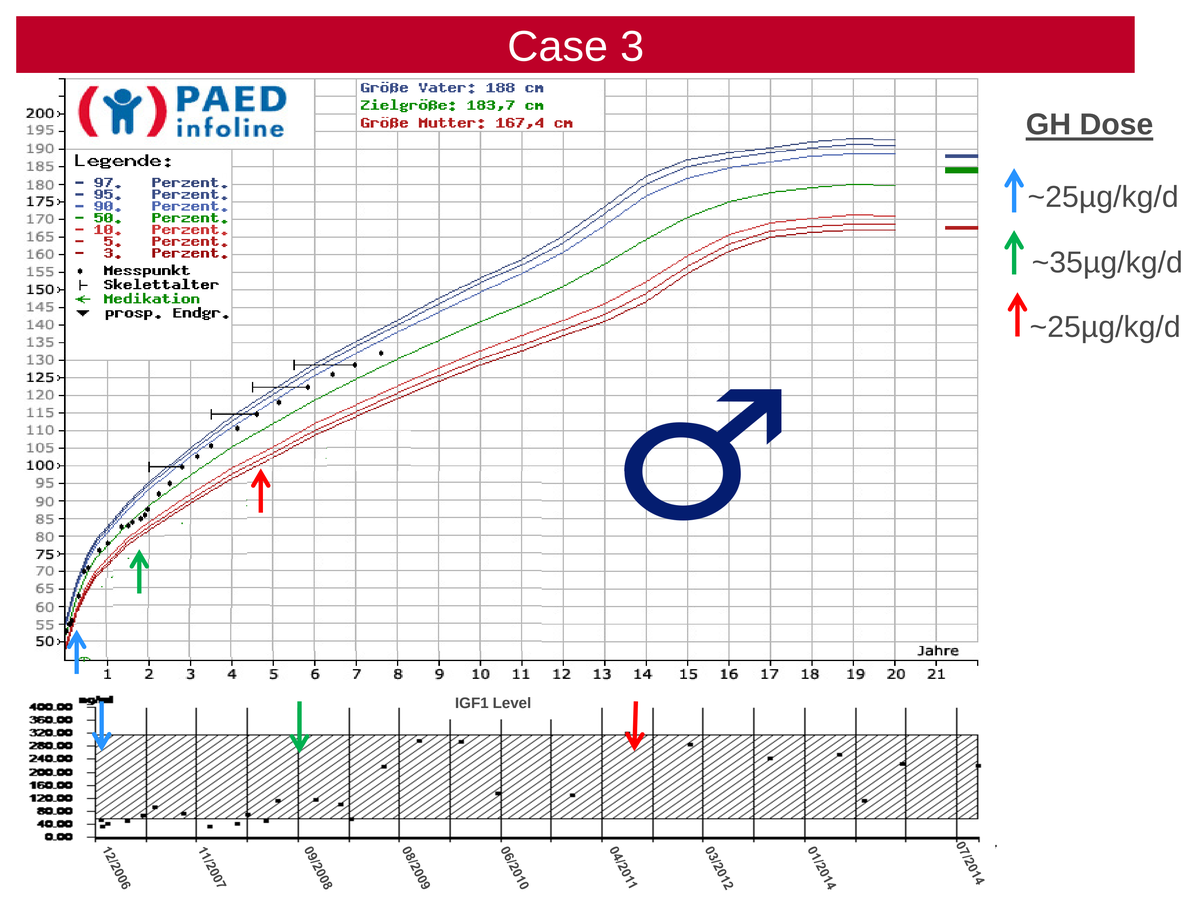
Case 1



Case 2



Case 3



Picture 1 shows growth charts and IGF1 levels during the treatment with GH. Time scale differs for better viewing conditions

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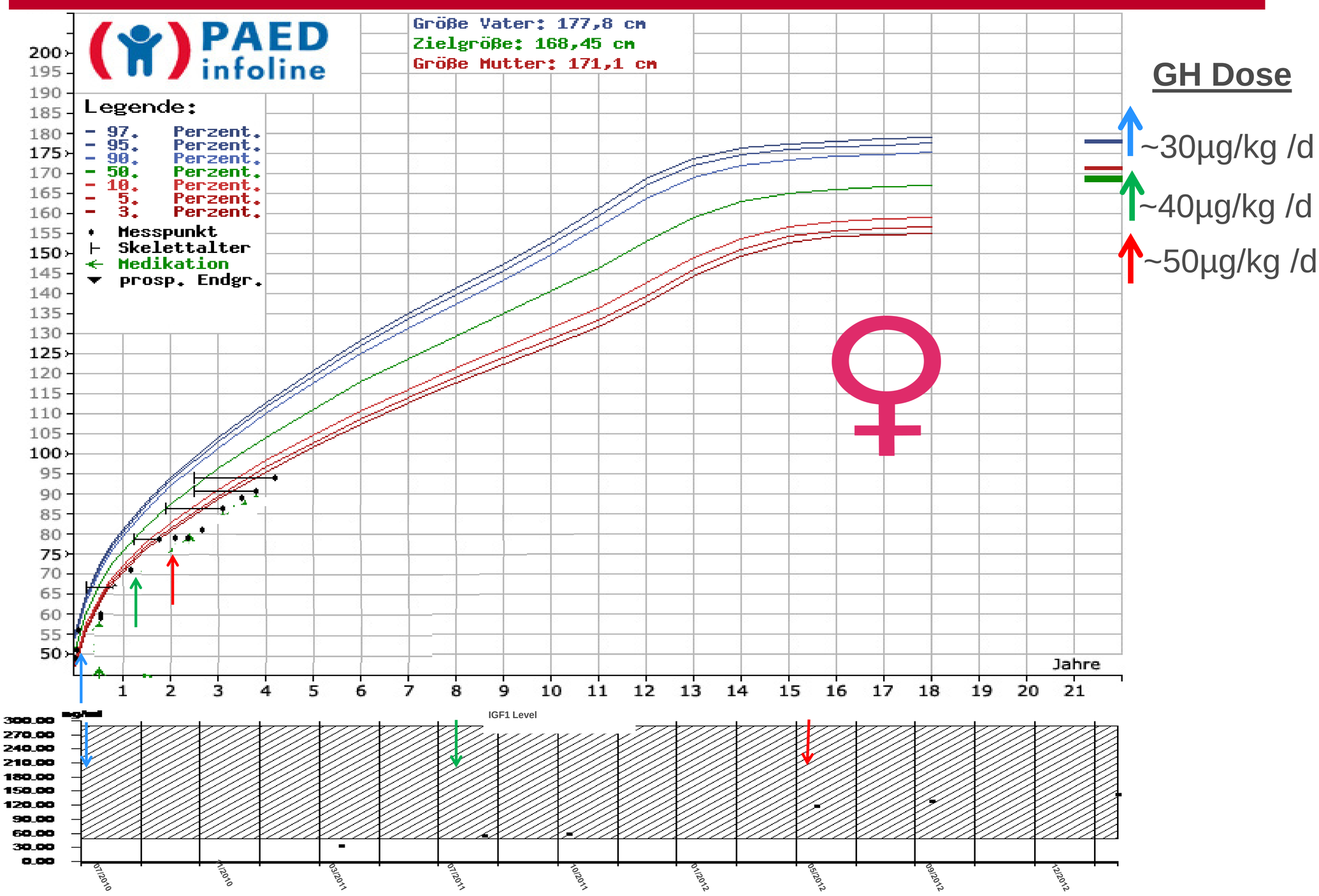

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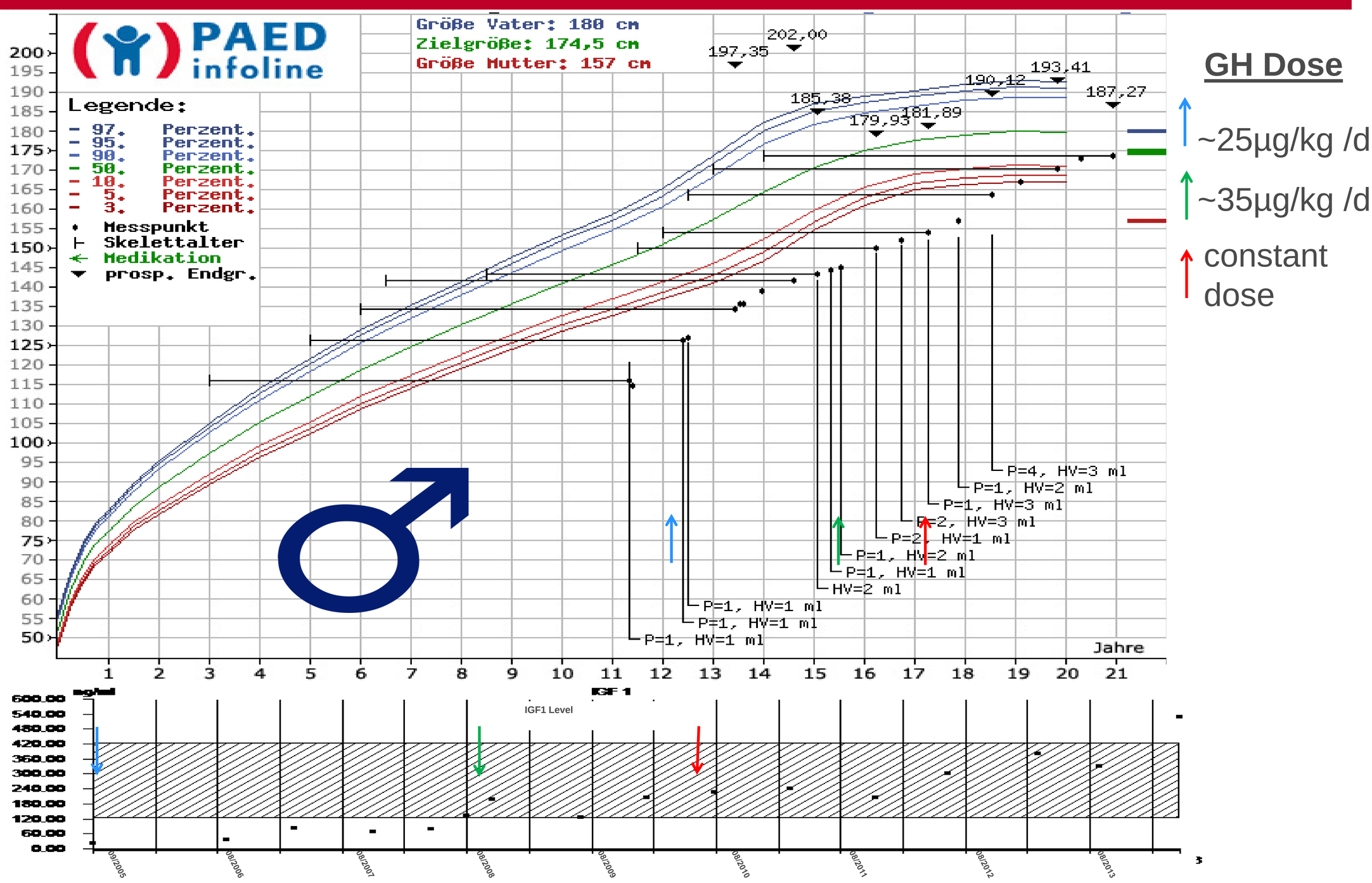
Case 2



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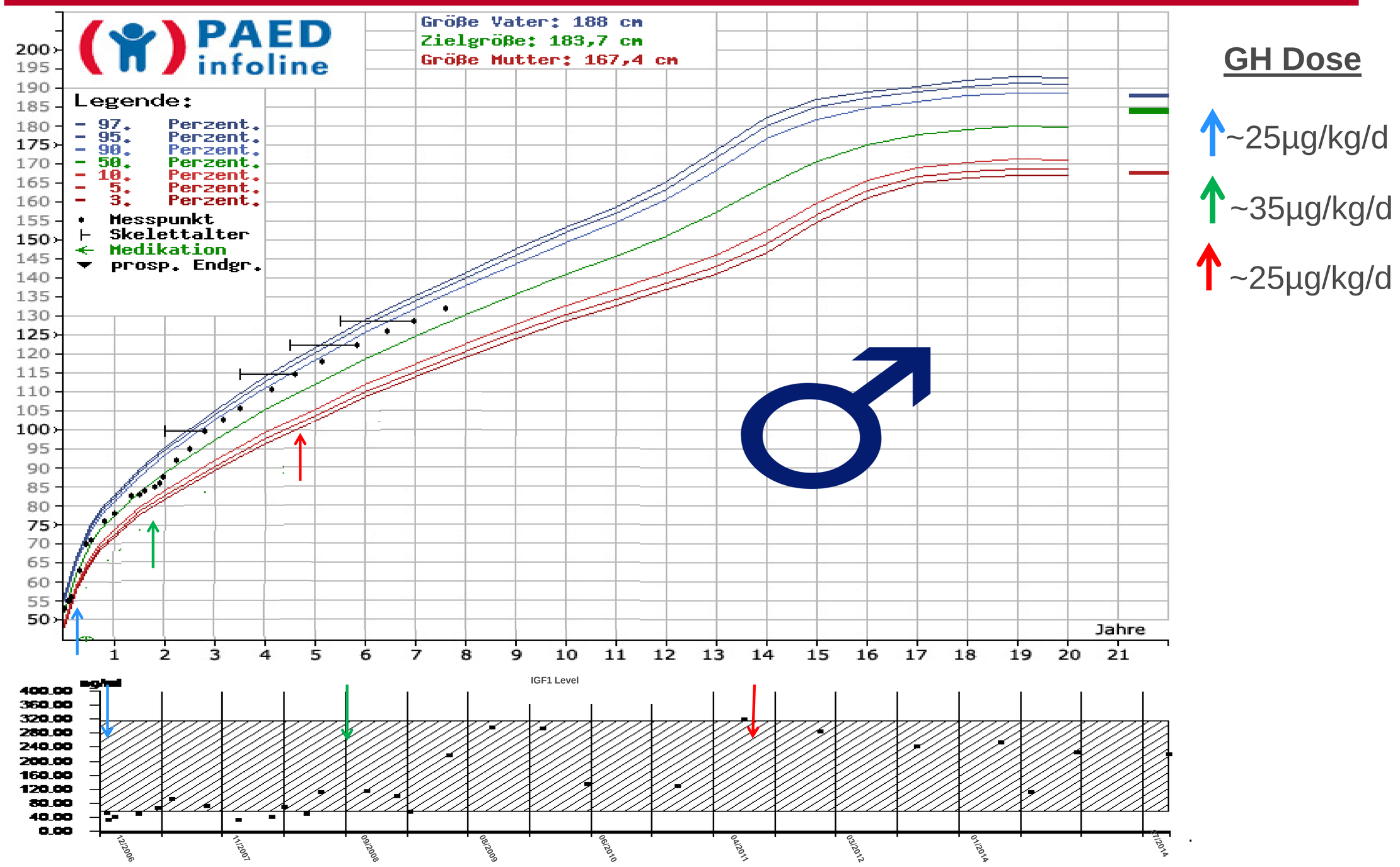

GH Dose

↑ ~25µg/kg/d

↑ ~35µg/kg/d

↑ ~25µg/kg/d

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