

# The influence of Growth Hormone treatment on the Basal Metabolism in Kabuki Syndrome children.

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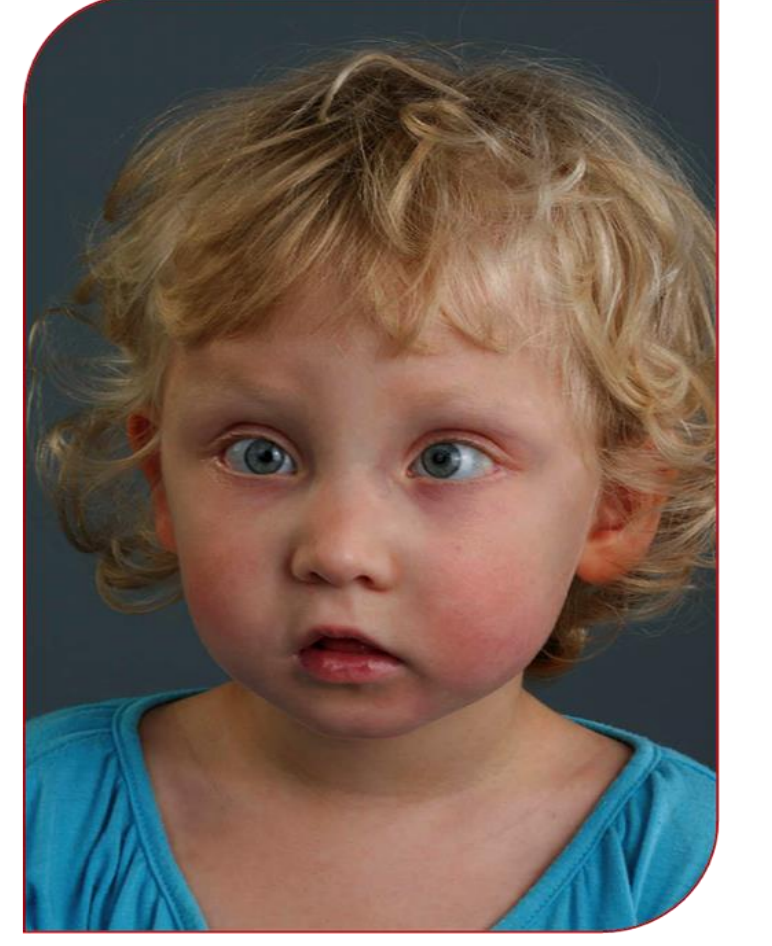
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## Introduction

The influence of growth hormone on the metabolism of prepubertal children with Kabuki syndrome was never investigated before. Kabuki syndrome is a rare syndrome, which is mainly characterized by mental retardation, short stature, specific facial features, obesity and hypotonia. This syndrome is caused by a mutation in the *KMT2D* or *KDM6A* genes. Studies in children with Prader-Willi syndrome, showed the effectiveness of growth hormone treatment on the basal metabolism<sup>A</sup>, we expected the same effect in children with Kabuki syndrome. In this prospective study we investigated the influence of growth hormone on the basal metabolic rate (BMR) and physical activity in genetically proven, prepubertal children with Kabuki syndrome.

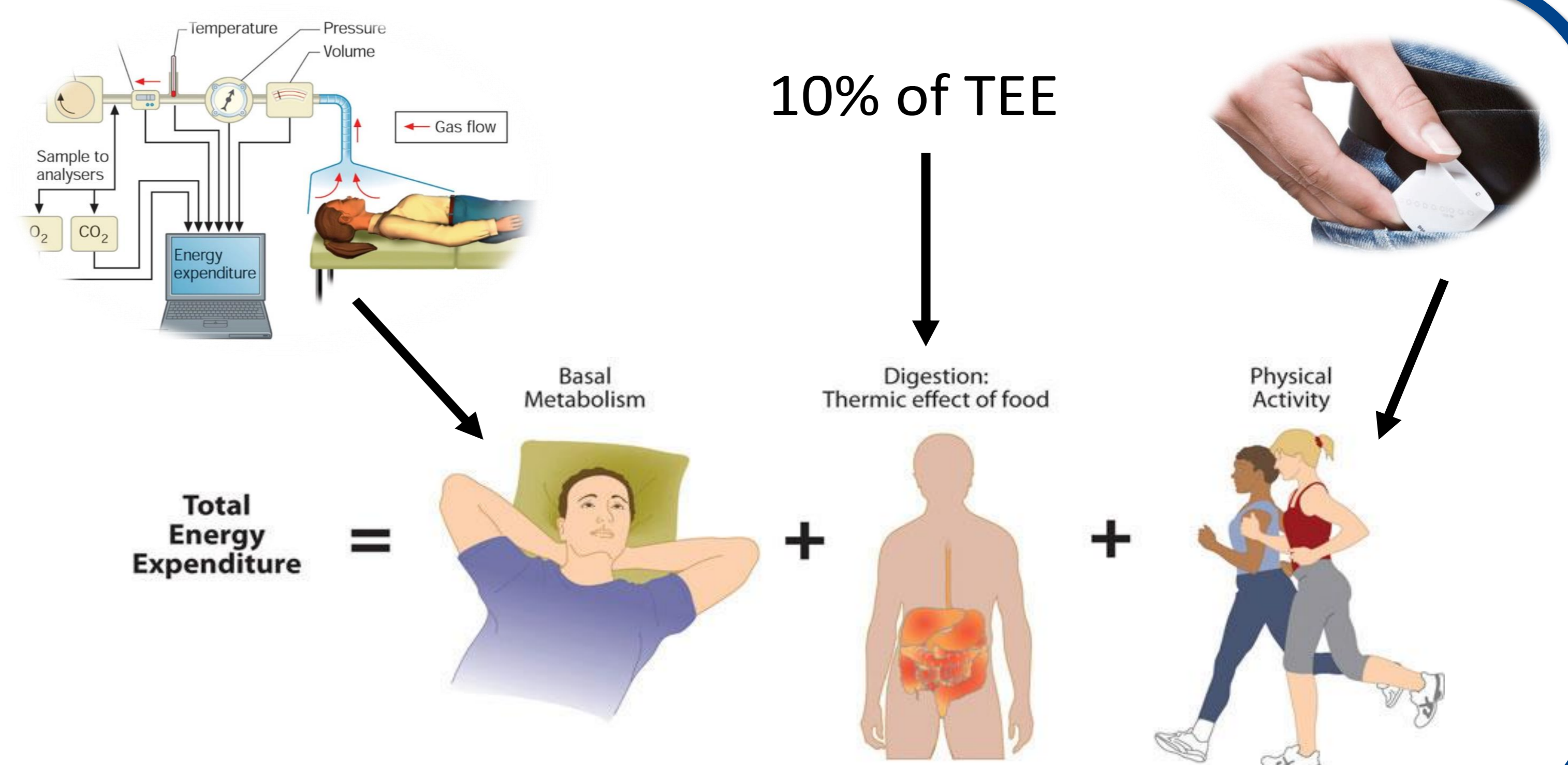


## Material & Methods

We included 17 children with Kabuki syndrome, 8 girls and 9 boys with a mean age of  $6.87 \pm 2.23$  year.

The BMR before start and during growth hormone treatment was measured with the ventilated hood (VH) technique. The BMR was calculated by Weirs formula<sup>B</sup>.

The accelerometer was used to measure the physical activity. We used the cut off point of at least 3 days wearing time with a minimum of 600 min/day. Ten of the 17 children met the criteria.



## Results

The BMR was significantly increased by growth hormone from  $3.17 \pm 0.52$  kJ/min to  $3.70 \pm 0.57$  kJ/min ( $p=0.001$ ). The physical activity was assessed by accelerometer. Six out of 10 children had an increased physical activity during growth hormone treatment compared to baseline (not significant).

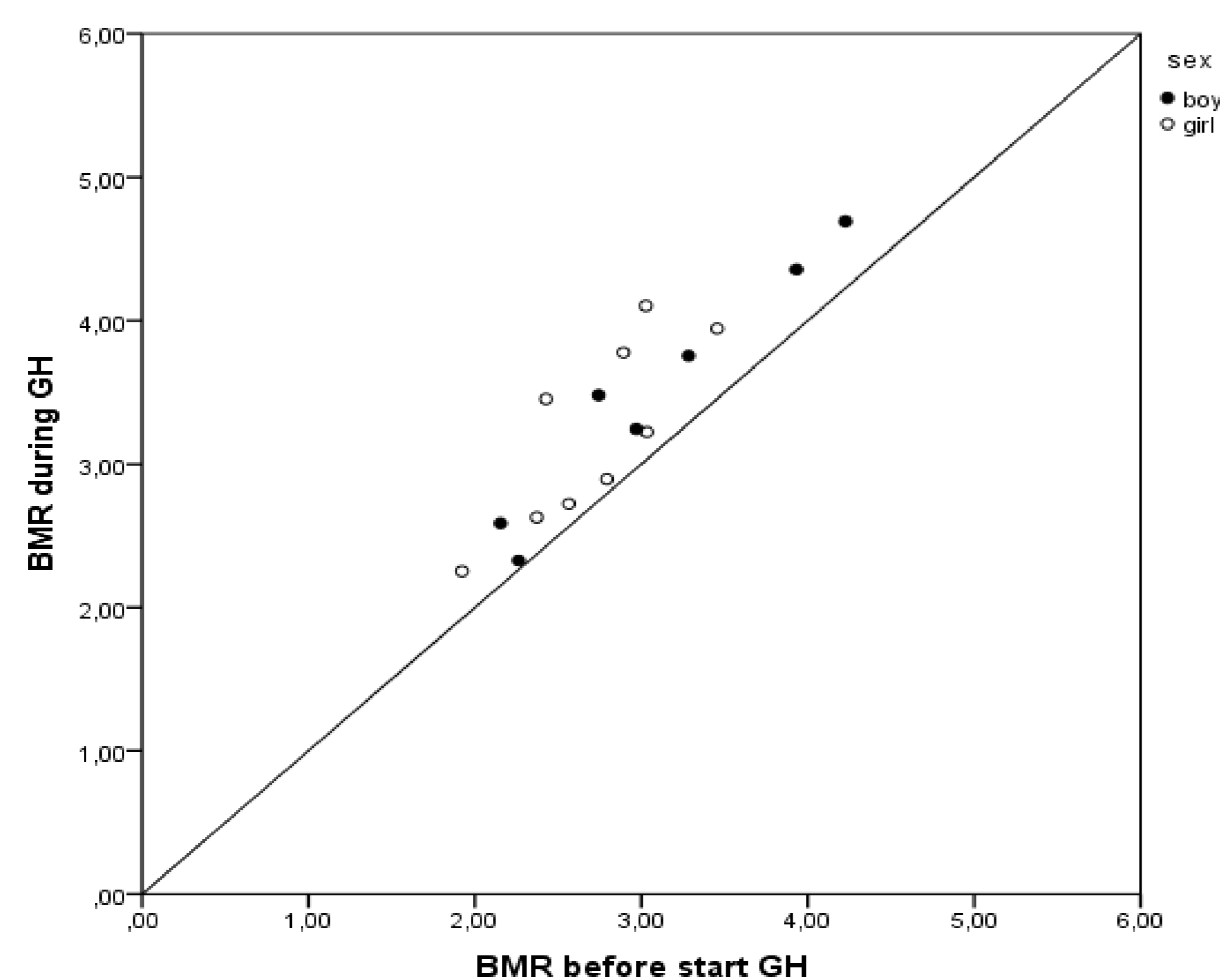


Figure 1: Growth hormone treatment significantly increased the BMR.

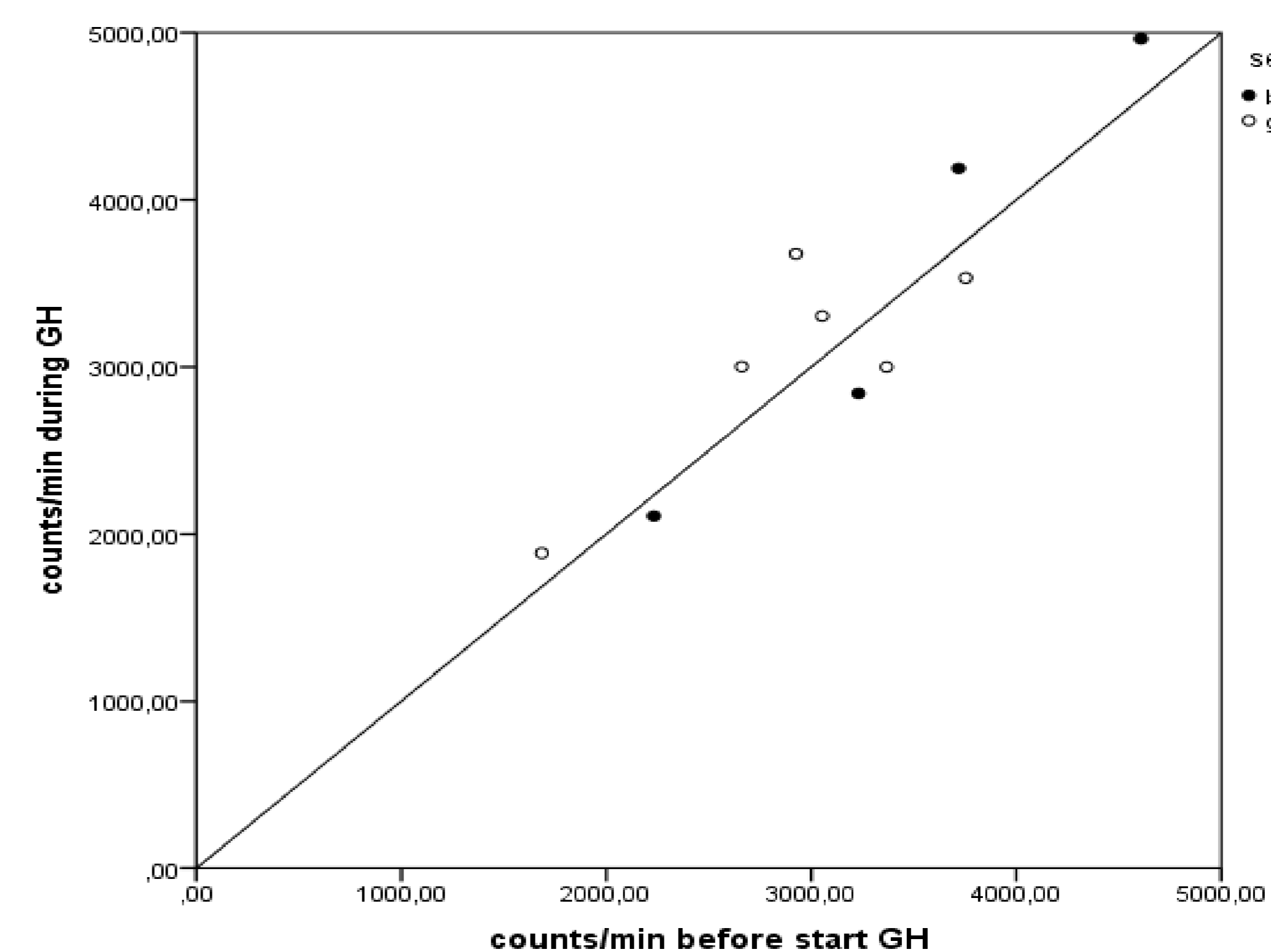


Figure 2: Physical activity is not significantly changed by growth hormone treatment.

## Conclusion

The BMR increased significantly during growth hormone treatment compared to before. The children who wore the accelerometer had a mean wearing time of 8.5 days and this resulted for 6 out of 10 children in a higher physical activity during growth hormone treatment than before the start of growth hormone (not significant). In conclusion, this suggests that the increased BMR is caused by the growth hormone treatment instead of an increased physical activity.

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B. Weir, J. D. V. New methods for calculating metabolic rate with special reference to protein metabolism. *The Journal of physiology*, 109. 1-2 (1949): 1.

