

Improved mental and motor development during GH treatment in very young children with Prader-Willi syndrome

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Conclusion

Mental and motor development increased significantly during 3 years of GH treatment.

Background

Infants and toddlers with Prader-Willi Syndrome (PWS) have a mental and motor developmental delay. Short-term data suggest a positive effect of growth hormone (GH) on mental and motor development. There are, however, no longer-term results about the effects of GH treatment on mental and motor development.

Aim

To investigate the longer-term effects of GH on psychomotor development in infants and toddlers with PWS and the effect of age at start of GH treatment on psychomotor development.

Participants & Method

Prospective cohort study in 63 young children with PWS. All patients were naïve to GH treatment at time of enrolment and started GH at a dose of 1 mg/m²/d (≈ 0.035 mg/kg/d).

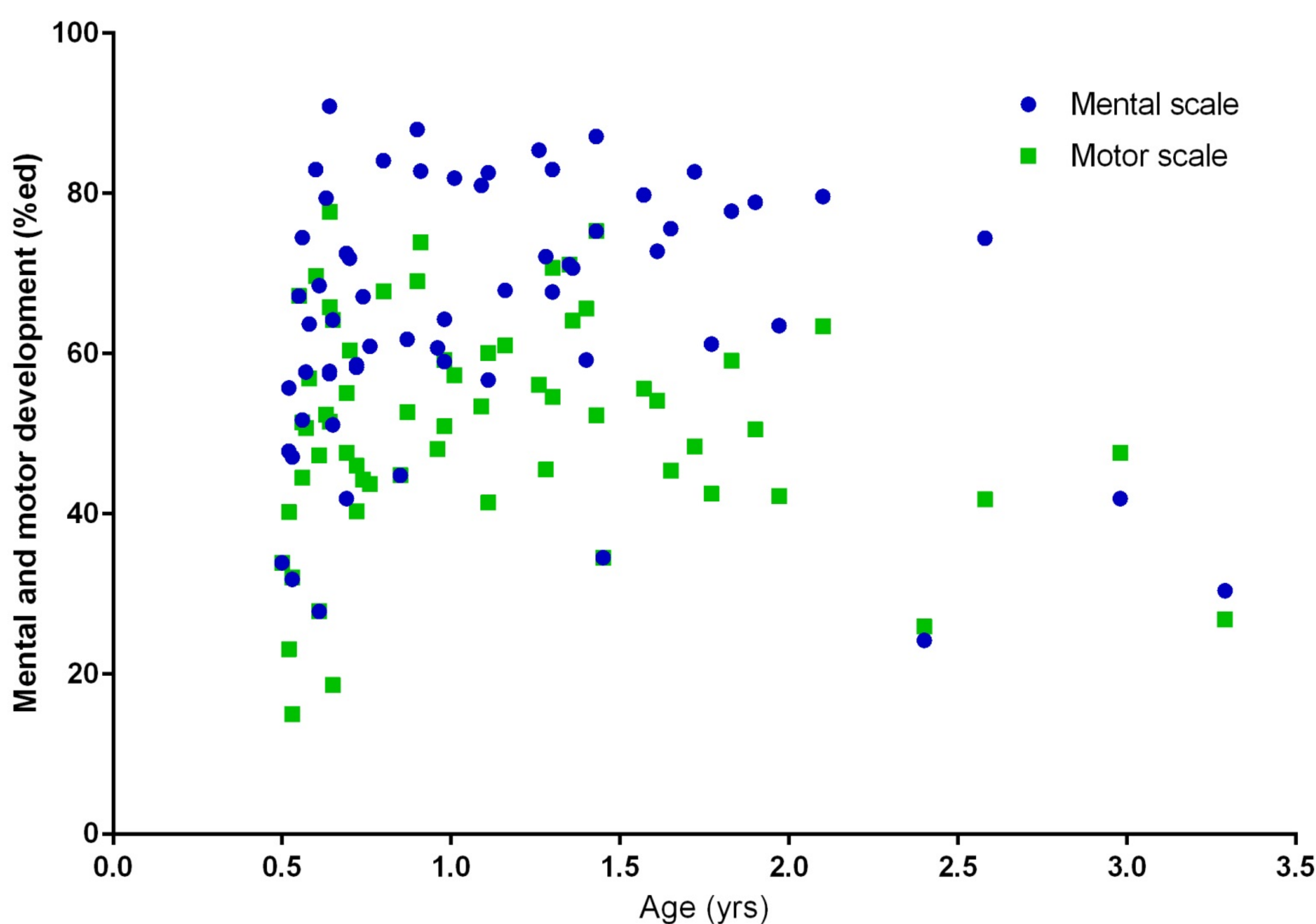
Main outcome measures were mental and motor developmental age assessed with Bayley Scales of Infant Development II (BSID-II) and expressed as % of the expected development (100%).

Baseline characteristics (n=63)

Age (years)	1.0	(0.7; 1.6)
Gender (♂ / ♀)	35 / 28	
Genetic subtype		
- Deletion	31 (49%)	
- mUPD	31 (49%)	
- Translocation	1 (2%)	
Height SDS	-1.7*	(-2.6; -1.0)
Weight for height SDS	-0.6*	(-1.4; 0.5)
Head circumference SDS	-1.3*	(-1.7; 0.5)
Fat mass percentage	28.6	(26.2; 34.6)

Data expressed as median (IQR). * p<0.01, compared to 0 SDS

Mental and motor development before start of GH



- During 3 years of GH, mean (SEM) mental development increased from 58.1% (2.8) to 79.6% (3.7) and motor development from 41.9% (2.9) to 78.2% (3.9; both p<0.01).

- A lower baseline psychomotor development and a younger age at start of GH were significantly associated with a higher increase in mental and motor development (p<0.01).

Psychomotor development during 3 years

