Growth outcome and metabolic profile of PWS patients treated with GH and differences between AGA and SGA group

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BACKGROUND & PURPOSE

Prader-Willi syndrome (PWS)

A complex genetic disease associated with growth impairment, severe obesity and metabolic dysfunctions.

High proportion of PWS patients are born small for gestational age (SGA), which also increase the risk of growth impairment and metabolic dysfunction

The aim of this study

Describe growth outcome and metabolic profiles in GH treated PWS patients.

Investigate the differences in clinical outcomes between adequate for gestational age (AGA) and SGA group

PATIENTS & METHODS

Inclusion

Genetically verified PWS patients diagnosed at Pusan National University Children's Hospital between 2008-2019

More than 2 years old

Treated with GH for more than 1 year

Exclusion

who reveived concurrent GnRHa treatment

Methods

Retrospective chart review

Comparison between SGA and AGA group (SPSS 12)

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RESULTS

Table 1. Patient characteristics

Variable	All patients (n=55)	SGA (n=20)	AGA (n=35)
Male (n, %)	32(58.2)	15 (75.0)	17 (48.6)
Current age (yr)	9.0±4.0	8.8±3.8	9.2±4.1
Age at GH initiation (yr)	2.2±2.6	1.6±1.8	2.5±2.9
Duration of GH treatment (yr)	6.3±3.0	7.0±2.6	5.9±3.0
Gestational age (weeks)	38.6±1.9	38.8±1.3	38.5±2.2
Birth weight (kg)	2.68±0.46	2.38±0.29	2.85±0.45
Genetic causes deletion (n, %) uniparental disomy (n, %)	 39 (70.9) 16 (29.1)	 13 (65.0) 7 (35.0)	 26 (74.3) 9 (25.7)

RESULTS

Table 2. Anthropometric characteristics and laboratory data

Variable	All patients (n=55)	SGA (n=20)	AGA (n=35)
Ht-SDS at GH initiation	-0.91±1.60	-1.23±1.26	-0.77±1.73
Current Ht-SDS	-0.29±1.34	-0.16±1.16	-0.36±1.44
BMI-SDS at GH initiation	1.64±1.97	1.33±2.42	1.78±1.85
Current BMI-SDS	1.40±1.19	1.33±1.20	1.44±1.21
HbA1c (%)	5.4±0.3	5.46±0.43	5.42±0.24
Glucose (mg/dL)	105.4±22.0	114.4±27.8	100.1±16.0
Triglyceride (mg/dL)	111.2±66.2	103.8±39.8	115.6±72.4
Total Cholesterol (mg/dL)	184.0±29.3	184.7±28.8	182.7±30.7
LDL (mg/dL)	105.9±25.2	104.6±24.3	106.6±26.0

Ht,height; SDS,standard deviation score; BMI,body mass index

Table 3. GH treatment effect and metabolic dysfunction

Variable	All patients (n=55)	SGA (n=20)	AGA (n=35)
Current Ht-SDS	-029±1.34	-0.16±1.16	-0.36±1.44
ΔHt-SDS(current – GH initiation)	0.66±1.50	1.05±1.32	0.50±1.56
ΔBMI-SDS (current – GH initiation)	-0.40±1.88	-0.55±3.03	-0.33±1.27
DM (n, %)	2 (3.6)	2 (10.0)	0
Overweight/obesity (n, %)	34 (61.8)	13 (68.4)	21 (65.6)
Dyslipidemia (n, %)	21 (38.2)	7 (35.0)	14 (40.0)

Ht,height; SDS,standard deviation score; BMI,body mass index; DM, diabetes mellitus

SUMMARY & CONCLUSIONS

In GH-treated Prader-Willi syndrome patients,
we compared growth outcome and metabolic profile between SGA and AGA group

- Anthropometric parameters did not differ between AGA and SGA group
- Growth hormone effect did not differ between AGA and SGA group
- Glucose level was higher in SGA group, and 2 SGA patient had DM

so more careful monitoring and prevention for DM will be required in SGA group