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Premature Pubarche in Prader-Willi Syndrome: Potential Predictors and Consequences

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Background/Objective

- Children with PWS may develop premature pubarche (PP) (1).
- We studied the incidence of PP and its potential precursors and consequences.

Methods:

- Retrospective chart review
- Inclusion criteria:
 - Children with PWS treated at Children's Mercy Hospital between 1990 – 2021
- Exclusion criteria:
 - Not seen in endocrine clinic
 - No documentation of pubarche
- Wilcoxon rank-sum, Fisher's exact, and Kendall rank correlation coefficient (CC) were used.

Table 1: Table of Definitions

Terminology	Definition
PP	Tanner stage 2 (TS2) pubic hair in girls < 8 and boys < 9 years old.
Gonadarche	TS2 breast development or testicular volume ≥4 mL.
Index for bone age advancement	Bone age (BA) divided by chronological age (CA) (BA/CA), with BA/CA > 1 being advanced
Homeostatic Model Assessment for Insulin Resistance (HOMA-IR)	Calculated as (Fasting insulin, uIU/mL)*(Fasting glucose, mg/dL)/405.
Small for gestational age (SGA)	Birth weight less than the 10% for age.
Type 2 Diabetes Mellitus (T2D)	HbA1c ≥6.5%.

Figure 1: Breakdown of patients with premature pubarche (PP) versus normal puberty (NP)

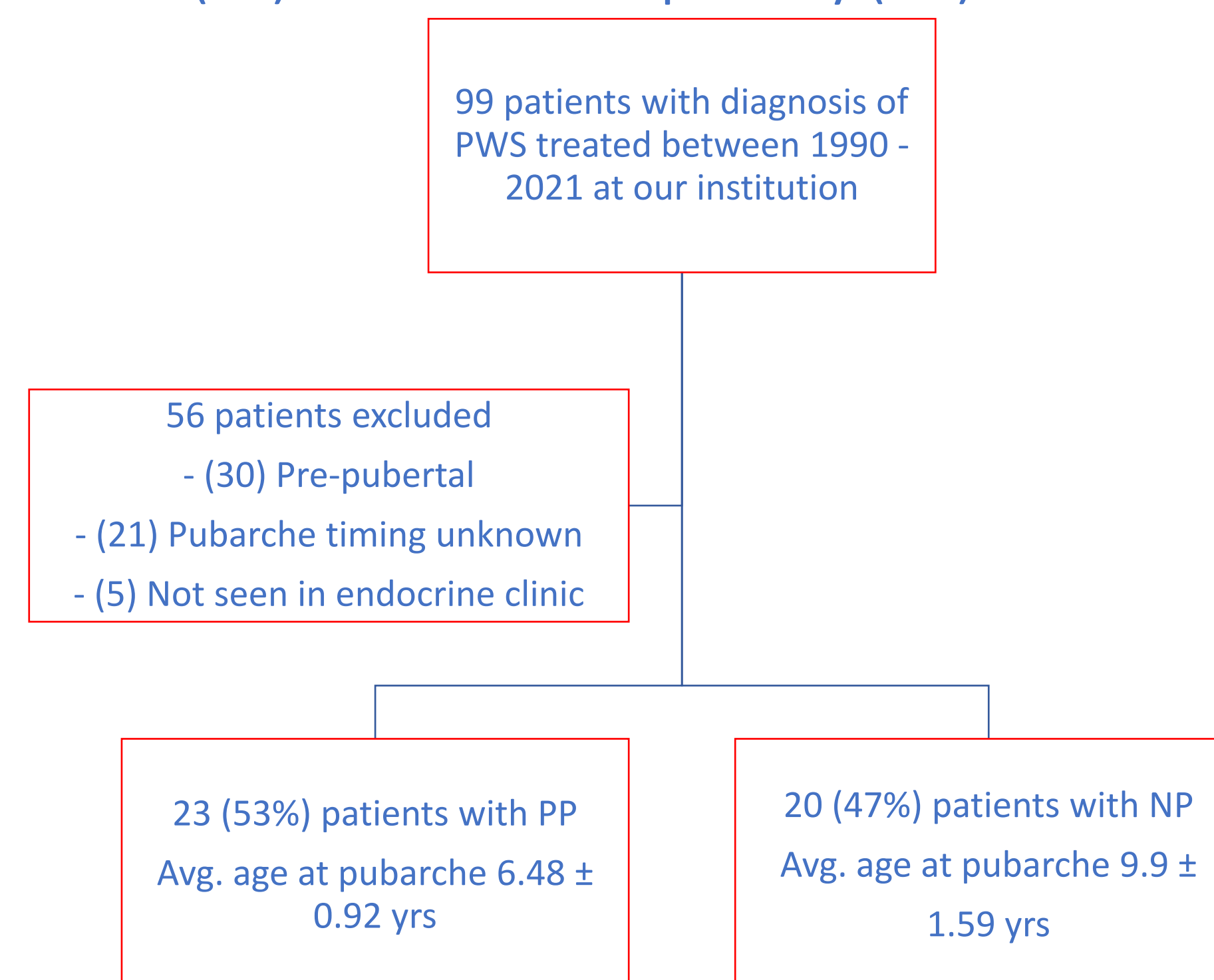


Table 2: Patient Characteristics

	Premature Pubarche	Normal Pubarche	p-value
Race			0.532
	Black 5 (21.7%)	3 (15.0%)	
	White 16 (69.6%)	17 (85.0%)	
	Multiracial 2 (8.7%)	0 (0.0%)	
Sex			0.227
	F 9 (39.1%)	12 (60.0%)	
	M 14 (60.9%)	8 (40.0%)	
SGA status			0.99
	No 15 (78.9%)	12 (75.0%)	
	Yes 4 (21.1%)	4 (25.0%)	
Age at gonadarche	9 [9.0, 11.0]	10.5 [10.0, 12.0]	0.207

Table 3: Correlation with Age at Pubarche

	N	Correlation	p-value
Gestational age	37	-0.08	0.52
BMI z-score	39	-0.12	0.31
HOMA-IR	17	0.16	0.42
rhGH initiation age	37	0.08	0.55

Results:

Table 4: Premature Pubarche and its relationship to final height and BA advancement at gonadarche and pubarche

	Premature Pubarche		Normal Pubarche		P-value
	N	Median [IQR]	N	Median [IQR]	
Difference between target height and final adult height	6	-13.3 [-20.0, 1.2]	13	-9.6 [-18.4, -3.3]	0.507
BA/CA at pubarche	15	1.1 [1.0, 1.4]	10	1.0 [1.0, 1.2]	0.210
BA/CA at gonadarche	10	1.2 [1.1, 1.3]	6	1.1 [1.0, 1.2]	0.175

Figure 2: Correlation between BMI z-score at pubarche and pubarchal BA advancement

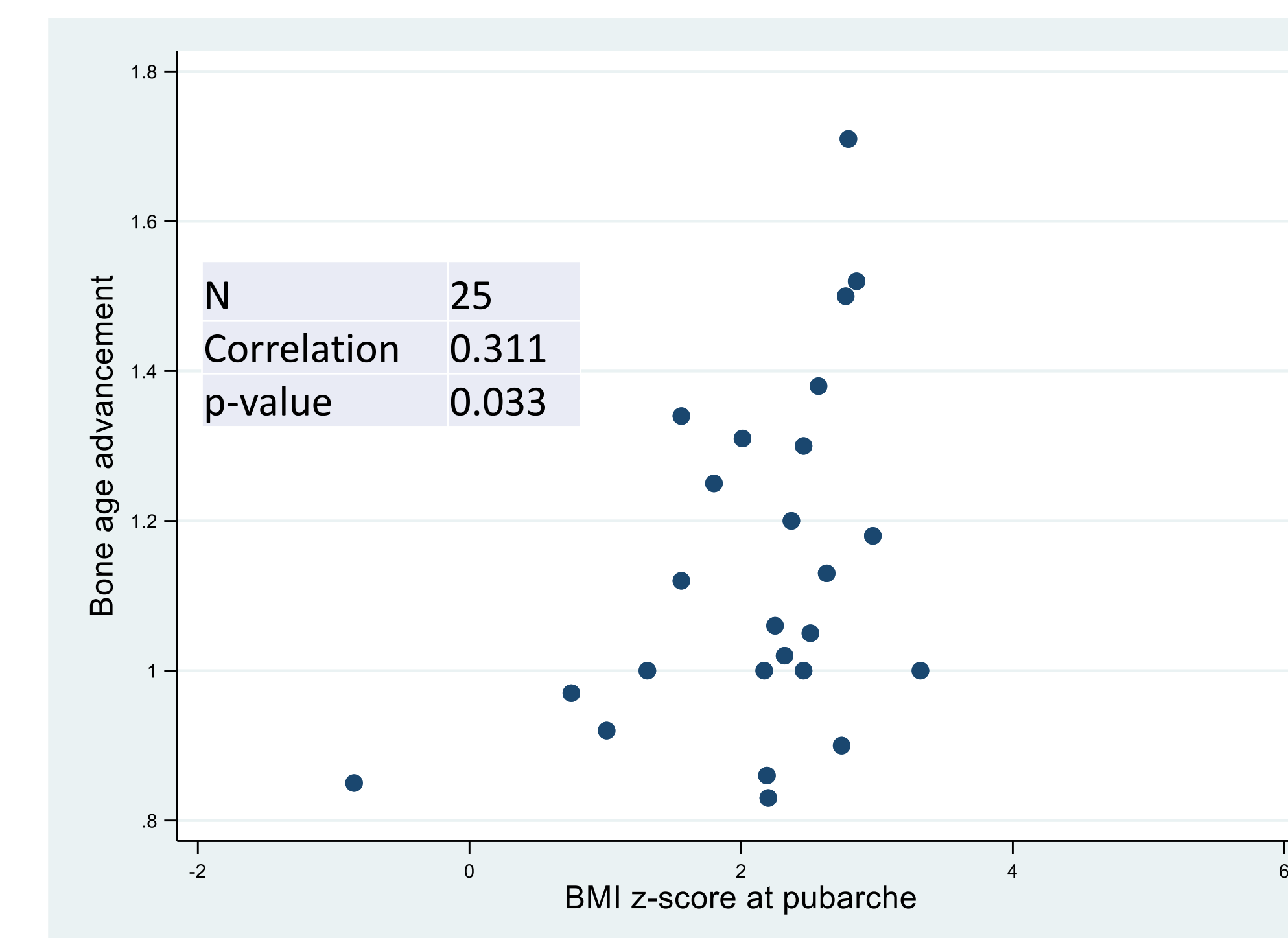


Table 5: Metabolic Consequences of Premature Pubarche

	Premature Pubarche		Normal Pubarche		P-value
	N	Median [IQR]	N	Median [IQR]	
Highest A1c after pubarche	20	5.8% [5.6, 6.1]	18	5.6 [5.4, 6.0]	0.356
Type 2 Diabetes after pubarche	21	4 (19.1%)	18	6 (33.3%)	0.465
Dyslipidemia after pubarche	21	18 (85.7%)	16	11 (68.7%)	0.254
Total cholesterol	20	190.5 [181.0, 215.0]	16	179.5 [154.0, 199.5]	0.092
HDL	20	40.5 [32.5, 54.0]	16	54.5 [42.0, 66.5]	0.041
LDL	20	123.0 [111.0, 149.5]	16	89.0 [75.5, 124.5]	0.107
Triglycerides	20	124.5 [84.5, 158.5]	15	99.0 [67.0, 158.0]	0.142

Conclusions:

- Premature pubarche is common in PWS but does not impact discrepancy from final height, though obesity at pubarche results in advanced bone age.
- Obesity and insulin resistance may not be the cause of premature pubarche in children with PWS, contrary to what has been seen in obese children without PWS (2).
- Patients with PP had significantly lower HDL and trends towards higher total cholesterol, LDL, and triglycerides.

References:

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