

# UNTREATED JUVENILE HYPOTHYROIDISM LEADING TO UNUSUAL PRESENTATION -VAN WYK GRUMBACH SYNDROME

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## INTRODUCTION

VAN WYK GRUMBACH SYNDROME [VWGS] is a rare complication of prolonged untreated juvenile hypothyroidism, characterised by high levels of TSH, isosexual precocious puberty, enlarged multicystic ovaries [Type 3 OHSS], pubertal uterus, enlarged pituitary in imaging and delayed growth and bone age.

It is the only form of precocious puberty in which bone age is delayed.

Phenotypically these girls show classical hypothyroid appearance, delayed growth, FSH mediated secondary sexual characteristics with breast development with or without galactorrhoea, uterine bleeding but absence of significant pubic or axillary hair development.

Here I present a case of VWGS where patient's findings were completely reversed with thyroxine replacement.

## CASE REPORT

A 15 year old girl was brought to our OPD with H/O stunted growth, oligomenorrhoea followed by menorrhagia since menarche, loss of appetite but increase in weight for the past 1 year. Her scholastic performance is average. On examination, patient looked pale with dry skin. On anthropometry, patient's height was 131.5 cms. Her Tanner stage was prepubertal [no axillary or pubic hair] except for thelarche. Her systemic examination was unremarkable. Visual acuity and fields were normal.

Laboratory testing showed Haemoglobin-10.5g/L, TSH >150 microIU/L, FT4-0.08ng/dL, FSH -8.18mIU/ml, LH-0.26mIU/ml, Prolactin-137ng/ml, Estradiol-610pg/ml.

Her radiological investigations revealed a bone age of 7. Ultrasonography of pelvis revealed uterus with pubertal size 8.9x4.6cm, right ovary shows a multiloculated cyst 8.9x7.1cms, left ovary shows a multiloculated cyst 8.4x6.2cms. MRI of sella revealed pituitary hyperplasia.

On the basis of clinical history, physical examination and additional investigations, patient was diagnosed as VWGS.

## RESULTS

The patient was started on thyroxine replacement 75micrograms/day, which was titrated by periodic lab investigations. During followup, her thyroid function and serum prolactin normalised at 6 months. Repeat USG revealed a complete regression of ovarian cysts, with normal sized ovaries at 6 months.

## DISCUSSION

VWGS is a rare presentation of untreated hypothyroidism. The prepubertal response in VWGS is always isosexual and mediated by very high TSH levels acting through the FSH receptors inducing a follicle stimulating hormonal effect. Early recognition and thyroid hormone replacement can completely regress symptoms and ovarian enlargement, while improving final height achievement.

Biochemically low free thyroxine [T4] is combined with raised levels of TSH, PRL and estradiol. Typically LHRH stimulation shows an FSH dominated prepubertal response with suppressed LH confirming GnRH independent precocious pseudopuberty.

## CONCLUSION

Hypothyroidism is a relatively common endocrine disorder among children and early recognition is important to prevent serious complications like VWGS. Sexual precocity with delayed bone age and stunting should create high clinical suspicion towards this unique diagnosis. As symptoms regress with thyroid hormone replacement, early diagnosis and timely adequate replacement of thyroxine can avoid unnecessary surgical interventions.

BEFORE TREATMENT

AFTER TREATMENT



## REFERENCES

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